SYMPOSIUM

DENTAL MANAGEMENT OF THE HANDICAPPED CHILD
SUPPORT

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CONTENTS

SYMPOSIUM

DENTAL MANAGEMENT OF THE HANDICAPPED CHILD

Program 1

Contributors 3

Gerald H. Prescott

Genetics and the Prevention of Mental Retardation 7

Hans Zellweger

Neuromuscular Diseases Involving Craniofacial Structures 15

Ronald Johnson

Hospital Dentistry for the Handicapped Child 25

Harold Turner

Anesthesia and Analgesia: Adjuncts in the Dental Management of the Handicapped Child 35

Rudy Hormuth

A New Direction in the Training of Professionals for Delivery of Services to the Handicapped 46

Arthur J. Nowak

Preventing Dental Disease in the Handicapped Child: A Challenge to the Professions 55

Osamu Chiono

A Comprehensive Clinical Dental Program for Hemophiliacs 67

Osamu Miyamoto

Organization of an Interdisciplinary Unit for the Study of Craniofacial Anomalies 81

Samuel Pruzansky

Future Care of the Handicapped Person 87

Richard Koch


PROGRAM

Symposium
Dental Management of the Handicapped Child

Tuesday, May 22, 1973

8:00- 8:35  Registration
8:35- 8:45  Introduction: Dr. C. A. Full
8:45-10:00 "Genetics and the Identification and Prevention of Inherited Diseases"
            Dr. G. H. Prescott
10:00-10:15 Moderator: Dr. W. Gibson
10:15-10:30 Coffee Break
10:30-11:45 "Neuromuscular Diseases Involving Craniofacial Structures"
            Dr. H. Zellweger
11:45-12:00 Moderator: Dr. W. Gibson
12:00- 1:30 Lunch
1:30- 2:45 "Hospital Dentistry for the Handicapped Child"
            Dr. R. Johnson
2:45- 3:00 Moderator: Dr. R. Runzo
3:00- 3:15 Coffee Break
3:15- 4:30 "Anesthesia and Analgesia: Adjuncts in the Dental Management of the Handicapped Child"
            Dr. H. Turner
4:30- 4:45 Moderator: Dr. R. Runzo
5:30- 7:00 Refreshments
7:00- Banquet
      Speaker: Dr. R. Hardin
      Speaker: Mr. R. Hormuth
Wednesday, May 23, 1973

8:45-10:00 "Preventing Dental Disease in the Handicapped Child: A Challenge to the Professions"
Dr. A. Nowak

10:00-10:15 Moderator: Dr. J. Gullikson

10:15-10:30 Coffee Break

10:30-11:45 "A Comprehensive Clinical Dental Program for Hemophiliacs"
Dr. O. Chiono (paper delivered by Dr. O. Miyamoto)

11:45-12:00 Moderator: Dr. J. Gullikson

12:00-1:30 Lunch

1:30-2:45 "Organization of an Interdisciplinary Unit for the Study of Craniofacial Anomalies"
Dr. S. Pruzansky

2:45-3:00 Moderator: Dr. R. Jordon

3:00-3:15 Coffee Break

3:15-4:30 "Future Care of the Handicapped Person"
Dr. R. Koch

4:30-4:45 Moderator: Dr. R. Jordon
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GENETICS AND THE PREVENTION OF MENTAL RETARDATION

Gerald H. Prescott, D.M.D.

As major breakthroughs continue to be made in the field of genetics, more and more causes of mental and physical disabilities are being determined. (See Table 1 for a classification of genetic diseases.) Due to increasingly sophisticated laboratory techniques, most of the breakthroughs are occurring in the diagnosis of chromosome aberrations and the biochemical errors of metabolism. It is noteworthy that diseases causing mental retardation are found in all three major classifications of genetic disease.

At one time the major tool of the medical geneticist involved only counseling after a family had already given birth to a defective child with a genetic disease. The recent use of amniocentesis to detect prenatal disorders has become a practical tool that allows the geneticist to diagnose disease before birth at a time when a family can elect to terminate the pregnancy if they so desire. Also, as carrier testing becomes more widespread among the adult population for such diseases as sickle cell anemia and Tay-Sachs disease, sophisticated, thorough and compassionate counseling must be utilized so unnecessary emotional harm will not occur to involved families.

According to Hecht and Lovrien (2) some of the principles of genetic counseling are as follows:
1. Genetic counseling is an integral part of patient management.
2. The responsibility for genetic counseling rests with the doctor caring for the patient. He should provide the counseling or make sure that it is done.
3. Genetic counseling must be based on an accurate diagnosis.
4. A family history in pedigree form should be obtained prior to counsel-
ing. Reported evidence of disease in relatives should be confirmed whenever this may affect the genetic counseling.

5. Health workers from other disciplines (i.e., public health nurse, social worker) may need to be involved to help in the diagnosis, counseling and treatment of the patient and his family.

6. Genetic counseling does not include making decisions for the family. Certain decisions are the family’s responsibility and right.

7. Genetic counseling involves concern for the emotional as well as the physical consequences of the disease in the family.

8. Genetic counseling should be initiated as early as possible after the birth of the child in whom hereditary disease is suspected.

Once the pedigree has been obtained the diagnosis and counseling should be planned in stages.

It must be stressed that a pedigree is necessary to help determine the manner in which a trait or disease may be segregating in a family. This is particularly essential in the monogenic diseases, as McKusick (1) has catalogued some 1,800 diseases and the list is growing yearly. (Please refer to Table 2 for a partial list of the more common monogenic disease entities.)

Prenatal Diagnosis

The recent success in culturing cells from amniotic fluid has made it possible to detect chromosome aberrations as well as a growing list of biochemical errors. (Please refer to Table 3 for a partial list of biochemical diseases detectable by analyzing cultured fetal cells for the specific enzyme defect involved.)

Indications for amniocentesis are:

1. Advanced maternal age—40 years or over.

2. A previous child with Down’s syndrome or any other chromosome disorder.

3. A parent who is a chromosome translocation carrier.

4. A previous child with a severe sex-linked recessively narrative disease such as Duchenne’s dystrophy.

5. A significant chance of having a child with one of approximately 40 or 50 biochemical diseases now amenable to prenatal diagnosis. (3)

The principal benefits of prenatal diagnosis are not only preventing the birth of mentally handicapped individuals, but relieving the anxiety of families which risk having children with genetic disease. Another benefit is an economic one: serious genetic disease generates an economic burden that has to be borne by the family and by the public. For example, it is estimated that 4,000 children with Down’s syndrome are born each year in the United States. Public health surveys estimate that institutional care for
each child costs approximately $180,000 to $250,000 for its lifetime. (4) Nearly half of these children are born to women more than 35 years of age. Prenatal diagnosis certainly should be considered for monitoring pregnancies for older women whose risk of having children with Down’s syndrome is greater. Prevention of this disorder can be of great medical and socio-economic benefit.

As in most technologic breakthroughs, the moral and ethical aspects of prenatal diagnosis of genetic disorders are not maturing as rapidly as other aspects. Those working in genetics must realize that each family views their genetic defect in a different way; some take it lightly and some are devastated by the knowledge. Certainly many successful people, who are clinically normal and are able to contribute to society, have chromosome aberrations. Reflection and caution are indicated in advising parents making decisions about family planning in these emotional situations. Families that have children with many of the common malformations incur a risk for succeeding pregnancies that they should be aware of. (Refer to Table 4 for these recurrence figures.)

As knowledge increases in the field of genetics, techniques such as amniocentesis must be applied with caution by an informed profession to enlighten the public. It is of utmost importance that each family be provided with the best information at hand so that the decision they make will be the best one for them.

References

Other references
<table>
<thead>
<tr>
<th>Type</th>
<th>Frequency</th>
<th>Recurrence Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>I. Monogenic Diseases</strong> (some 1,800 entities according to McKusick)</td>
<td>1/200 Live Births</td>
<td>High (25-50%)</td>
</tr>
<tr>
<td>a. Autosomal dominant</td>
<td></td>
<td></td>
</tr>
<tr>
<td>b. Autosomal recessive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>c. X-linked dominant</td>
<td></td>
<td></td>
</tr>
<tr>
<td>d. X-linked recessive</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>II. Chromosomal Aberrations</strong></td>
<td>1/250 Live Births</td>
<td>Variable (1-100%)</td>
</tr>
<tr>
<td>a. Those due to variation in number (Down's Syndrome, Turner's Syndrome)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>b. Those due to variation in structure (cat cry syndrome, 18q-, 4p-)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>III. Multifactorial Diseases</strong> (congenital malformation—neural tube defects, facial clefting, pyloric stenosis, ect.)</td>
<td>1/50 Live Births</td>
<td>Low (4-6%)</td>
</tr>
</tbody>
</table>

Table 2

**Monogenic Diseases**

<table>
<thead>
<tr>
<th>A. <em>Autosomal dominant</em></th>
</tr>
</thead>
<tbody>
<tr>
<td>Achondroplasia</td>
</tr>
<tr>
<td>Acrocephalosyndactyly: Apert's disease</td>
</tr>
<tr>
<td>Angionneurotic edema</td>
</tr>
<tr>
<td>Aniridia</td>
</tr>
<tr>
<td>Anonychia-ectrodactyly</td>
</tr>
<tr>
<td>Antitrypsin deficiency</td>
</tr>
<tr>
<td>Auriculo-osteodysplasia</td>
</tr>
<tr>
<td>Basal cell nevus syndrome</td>
</tr>
<tr>
<td>Brachydactyly</td>
</tr>
<tr>
<td>Camptodactyly</td>
</tr>
<tr>
<td>Cataract (some genetic forms)</td>
</tr>
<tr>
<td>Cleft lip and/or palate with lip pits</td>
</tr>
<tr>
<td>Cleft lip and/or palate with popliteal web</td>
</tr>
<tr>
<td>Cleidocraniodysostosis</td>
</tr>
<tr>
<td>Coproporphyria</td>
</tr>
</tbody>
</table>
Milroy's disease
Mandibulofacial dysostosis:
  Treacher Collins' diseases
Marfan syndrome
Moebius syndrome:
  congenital facial paralysis
Myotonia congenita:
  Thomsen's disease
Nail-patella syndrome
Nemaline myopathy
Oculodentodigital dysplasia
Ophthalmomandibulomalic dysplasia
Ophthalmoplegia, familial static
Optic atrophy, congenital
Osteogenesis imperfecta
Osteopetrosis:
  Albers-Schonberg disease
Osteopiklosis
Pachyonychia congenita
Paramyotonia congenita of Eulenberg
Polycystic
Porphyria
Ptosis
Pupillary membrane, persistent
Radioulnar synostosis
Retinal aplasia
Retinobastoma
Rieger's syndrome:
  dysgenesis of iris and cornea,
  hypodantia (and myotonic dystrophy)
Spherocytosis, hereditary
Split-hand deformity:
  lobster-claw deformity
Spondylopiophysseal dysplasia, congenital
Supravalvular aortic stenosis
Symphalagianism
Syndactyly
Triphalangeal thumb
Von Hippel-Lindau syndrome
Von Willebrand's disease:
  pseudo-hemophilia
Waardenburg's syndrome:
  white forelock, heterochromia iridis, and deafness
White sponge nevus of the mouth

B. Autosomal recessive

Abdominal muscles, absence of, with urogenital malformation
Abetalipoproteinemia
Acatalasemia
Adrenal hyperplasia
Albinism
Alkaptonuria
Alpers' diffuse degeneration of cerebral gray matter
AMAurosis congenita of Leber I
Aminoacidurias
Amyotonia congenita
Analphabetoproteinemia: Tangier disease
Anemia, congenital hypoplastic
Anophthalmos, true or primary
Antitrypsin deficiency of plasma
Aplasia cutis congenita
Argininosuccinic aciduria
Arterial calcification, generalized, of infancy
Asphyxiating thoracic dystrophy of the newborn
Ataxia-telangiectasia
Atrichia with papular lesions
Behr's infantile hereditary optic atrophy
Bloom's syndrome: dwarfism with skin changes
Cataract, congenital or juvenile
Chondrodystrophia calcificans congenita
Citrullinuria
Cockayne's syndrome: dwarfism, marble epiphyses, etc.
Corneal dystrophy, band-shaped
Cretinism, athyreotic
Crigler-Najjar syndrome:
  nonhemolytic jaundice
Cystathioninuria
Cystic fibrosis of the pancreas
Cystinuria, types I, II, and III
Dandy-Walker syndrome
Deaf-mutism and functional heart disease
Deaf-mutism (some types)
Diastrophic dwarfism
Disaccharide intolerance
Ellis-Van Creveld syndrome
Epidermolysis bullosa, letalis
Factor X deficiency
Fanconi’s pancytopenia
Fibrin-stabilizing factor deficiency
Fructose intolerance, hereditary
Galactosemia
Gangliosidosis, generalized
Gaucher’s disease, type II
Glancoma, congenital
Glutathione deficiency of erythrocytes
Glutathione reductase
Glycopenaemia
Glycogen storage disease, types I and II
Hallermann-Streiff syndrome
Heart-block, congenital
Hemochromatosis, idiopathic neonatal, giant cell hepatitis
Hexokinase deficiency hemolytic anemia
Histidinemia
Holoprosencephaly, familial alobar
Homocystinuria
Hurler’s syndrome
Hyperglycinemia, isolated
Hyperlipoproteinemia, type I
Hyperlipoproteinemia, type V
Hyperlysinemia
Hyperprolinemia, types I and II
Hypoglycemia, leucine-induced
Hypophosphatasia
Hypoprostaglandinemia
Hypotrichosis
Ichthyosiform erythroderma
Ichthyosis congenita
Isovalericacidemia
Krabbe’s disease
Larsen’s syndrome
Laurence-Moon-Biedl-Bardet syndrome
Leprechaunism
Lymphopenic hypergammaglobulinemia
Macular degeneration of the retina
Maple syrup urine disease
Marinesco-Sjörgen syndrome
Morquio syndrome
Mesomelic dwarfism of the hypoplastic ulna, fibula and mandible type
Metatrophic dwarfism
Methemoglobinemia
Microcephaly
Muscular atrophy, infantile
Nephrosis, congenital
Niemann-Pick disease
Oroticaciduria
Osteopetrosis
Phenylketonuria
Polycystic kidney, infantile, type I
Porphyria, congenital
PTA deficiency
Pycnodysostosis
Pyridoxine dependency
Pyruvate kinase deficiency
Reticulosis, familial histocytic
Retinitis pigmentosa
Rthmund-Thomson syndrome
Spastic diplegia, infantile type
Spastic paraplegia, hereditary
Spongy degeneration of central nervous system
Suxamethonium sensitivity
Tay-Sachs disease
Thalassemias
Usher’s syndrome
Weill-Marchesani syndrome
Wolman’s disease
Xeroderma pigmentosum

C. X-linked

Agammaglobulinemia, Swiss type
Albinism, ocular
Albinism-deafness syndrome
Aldrich’s syndrome of thrombocytopenia, infection and eczema
Angiokeratoma, diffuse: Fabry’s disease
Cataract, congenital, total

Cataract, congenital, with microcornea
Deafness, congenital, perceptive type
Diabetes insipidus, nephrogenic
Ectodermal dysplasia, anhidrotic
Glucose-6-phosphate dehydrogenase deficiency
Granulomatous disease, chronic
Hemophilia A
Hemophilia B
Hypopoposphatemic vitamin-D-resistant rickets
Ichthyosis
Iris, hypoplasia of, with glaucoma
Lesch-Nyhan syndrome
Lowe's oculo-cerebro-renal syndrome
Megalocornea

Microphthlamia or anophthalmia with digital anomalies
Mucopolysaccharidosis type II:
Hunter's syndrome
Ophthalmoplegia, external, and myopia
Retinos chisis
Van den Bosch syndrome: choroideremia, anhydrosis skeletal deformity and mental retardation

Table 3

Genetic Diseases Detectable by Studies of Fetal Cells in Amniotic Fluid

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Defective Enzyme or Metabolic Arrangement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chromosome abnormalities</td>
<td>Excess or deficiency of chromosomal material</td>
</tr>
<tr>
<td>Adrenogenital syndrome</td>
<td>Increased amniotic fluid corticosteroids</td>
</tr>
<tr>
<td>Arginosuccinic aciduria</td>
<td>Arginosuccinase</td>
</tr>
<tr>
<td>Citrullinemia</td>
<td>Arginosuccinic synthetase</td>
</tr>
<tr>
<td>Cystathioninuria</td>
<td>Cystathionase</td>
</tr>
<tr>
<td>Fabry's disease</td>
<td>Alpha galactosidase</td>
</tr>
<tr>
<td>Alpha fucosidosis</td>
<td>Alpha fucosidase</td>
</tr>
<tr>
<td>Galactosemia</td>
<td>Galactose-1-phosphate uridyl transferase</td>
</tr>
<tr>
<td>Gaucher's disease</td>
<td>Absent cerebrosidase</td>
</tr>
<tr>
<td>infantile type</td>
<td>Deficient cerebrosidase</td>
</tr>
<tr>
<td>adult type</td>
<td>Absent beta galactosidase</td>
</tr>
<tr>
<td>Generalized gangliosidosis</td>
<td>Cystathionine synthase</td>
</tr>
<tr>
<td>Homocystinuria</td>
<td>Valine transaminase</td>
</tr>
<tr>
<td>Hypervalinemia</td>
<td>Deficient beta galactosidase</td>
</tr>
<tr>
<td>Juvenile G_M1 gangliosidosis</td>
<td>Deficient hexosaminidase A</td>
</tr>
<tr>
<td>Juvenile G_M2 gangliosidosis</td>
<td>Alpha-I, 4 glucosidase</td>
</tr>
<tr>
<td>Glycogen storage disease type II</td>
<td>Multiple lysosomal hydrolases</td>
</tr>
<tr>
<td>&quot;I-cell&quot; disease</td>
<td>Isovaleryl CoA dehydrogenase</td>
</tr>
<tr>
<td>Isovaleric acidemia</td>
<td>Hypoxanthine-guanine phosphoribosyl transferase</td>
</tr>
<tr>
<td>Lesch-Nyhan syndrome</td>
<td>Alpha ketoisocaproate decarboxylase</td>
</tr>
<tr>
<td>Maple syrup urine disease</td>
<td>Absent arylsulfatase A</td>
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<td>Metachromatic leukodystrophy</td>
<td>Deficient arylsulfatase A</td>
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<td>late infantile type</td>
<td>Methylmalonyl CoA carboxylmutase</td>
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<td>juvenile and adult types</td>
<td>Sphingomyelinase</td>
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<td>Methylmalonic acidemia</td>
<td>Orotidyl pyrophosphorylase and orotidyl pyrophosphate uridyl transferase</td>
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<tr>
<td>Niemann-Pick disease</td>
<td>Phytoic acid alpha oxidase</td>
</tr>
<tr>
<td>Orotic aciduria</td>
<td>Hexosaminidase A and B</td>
</tr>
<tr>
<td>Refsum's disease</td>
<td>Increased amniotic fluid heparitin sulfate</td>
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<tr>
<td>Sandhoff's disease</td>
<td>Hexosaminidase A</td>
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<td>Sanfilippo's syndrome</td>
<td>Acid lipase</td>
</tr>
<tr>
<td>Tay-Sach's disease</td>
<td></td>
</tr>
<tr>
<td>Wolman's disease</td>
<td></td>
</tr>
</tbody>
</table>
### Table 4

Counseling Figures for Some Common Malformations

<table>
<thead>
<tr>
<th>Anencephaly or Meningo-Myelecele</th>
<th>Cleft Lip Plus or Minus Cleft Palate Alone</th>
<th>Pyloric Stenosis</th>
<th>Congenital Dislocation of Hip</th>
<th>Clubfoot</th>
</tr>
</thead>
<tbody>
<tr>
<td>Approximate incidence in general Caucasian population</td>
<td>2/1,000</td>
<td>1/1,000</td>
<td>1/2,500</td>
<td>2/1,000</td>
</tr>
<tr>
<td>Incidence in siblings of an affected child with unaffected parents</td>
<td>3.4-4.8/100</td>
<td>4.9/100</td>
<td>2.0/100</td>
<td>3.2/100</td>
</tr>
<tr>
<td>(brothers 4.0, sisters 2.4)</td>
<td></td>
<td></td>
<td></td>
<td>(brothers 0.5, sisters 6.3)</td>
</tr>
<tr>
<td>Incidence in children of an affected parent</td>
<td>4.3/100</td>
<td>6.0/100</td>
<td></td>
<td>Affected father 4.6</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Affected mother 16.2</td>
</tr>
</tbody>
</table>

Modified from Smith, D. W., and Aase, J. M. "Polygenic Inheritance of Certain Common Malformations: Evidence and Empiric Recurrence Risk Data." *J. Pediat.*, 76:653, 1970. These data are applicable for these anomalies as isolated malformations, but not when they occur as a part of a multiple malformation syndrome.
NEUROMUSCULAR DISEASES INVOLVING CRANIOFACIAL STRUCTURES

Hans Zellweger, M.D.

Diversified neuromuscular diseases affect craniofacial structures, or the muscles innervated by cranial nerves. In some conditions, the muscles are affected early in the course of the disease and may be the presenting symptoms to an extent that the mere inspection of the face allows one to recognize or at least suspect a particular neuromuscular disease. Dentists and other specialists (notably oral surgeons, otorhinolaryngologists and ophthalmologists) concerned with organs of the head and neck may be the first ones to see such patients, and thus it may be to their advantage to be familiar with some of these conditions.

It is the purpose of the first part of this presentation to discuss some basic principles of neuromuscular diseases, while in a second part a few specific diseases where involvement of facial, ocular and/or pharyngeal muscles prevails will be mentioned.

What is understood by the term neuromuscular diseases? These are diseases, most often genetic in nature, involving parts of the motor unit. The motor unit, by definition, originates in the ganglion cells of the anterior horns of the spinal cord or in the ganglion cells of the motor nuclei of the brain stem. It comprises, furthermore, the motor root, the peripheral motor nerve, the neuromuscular synapsis and, finally, the muscle fiber itself. As to localization of the disease process within the motor unit, the following categories of neuromuscular diseases can be distinguished:

1. diseases of the motor ganglion cells, e.g., spinal muscular atrophies;
2. diseases of the motor root and motor nerve, also called neuropathies,
for instance, the Guillain-Barre syndrome, which is not hereditary, and the Charcot-Marie-Tooth disease, or peroneal muscular atrophy, which is hereditary;
3. diseases of the neuromuscular synapsis, notably myasthenia gravis and myasthenia levior;
4. diseases of the muscle fiber, which include a great number of myopathies.

Clinical manifestations common to the neuromuscular diseases are muscular weakness (paresis), muscular hypotonia and muscle wasting (atrophy), disappearance of the tendon reflexes (hypo- or areflexia) and development of contractures. The latter develop if the weakness of the protagonists and antagonists attains different degrees. Weakness is defined as the decreased functioning of the volitionally (alpha fiber) innervated muscle, while hypotonia is defined as decreased resistance of the passive, not volitionally innervated muscle (muscle tone is under the control of the gamma system, which in turn is regulated by sensory input and diversified supranuclear centers). Muscle wasting or atrophy, i.e., decrease of muscle mass, is due to shrinking of the individual muscle fibers. The existence of nerve fibers exerting atrophic effect on the muscle has been well documented by various animal experiments. Muscle wasting is usually more pronounced in diseases of ganglion cells and motor nerves and appears later and to a lesser degree in myopathies.

Besides these manifestations common to all neuromuscular diseases, there are differences as to site of muscle weakness (facial muscles, pelvic and shoulder girdle muscles, distal muscles, etc.) and the speed of its development. In some conditions, severe generalized weakness and complete infirmity develop within a few months (infantile spinal muscular atrophy) or a few years (Duchenne muscular dystrophy), while other neuromuscular diseases show a slow course with little progression and barely affect life expectancy.

In diseases where the pathology sites are in the ganglion cells, fasciculations, a kind of wavy tremulousness, can be noticed. It is best seen in the tongue and in the intrinsic muscles of the hands, notably thenar and hypothenar muscles. In the neuropathies, sensory nerves participate often in the disease process and alterations of various superficial sensory and proprioceptive qualities are discovered. Sensory dysfunctions and fasciculations do not occur in the myopathies. Weakness due to diseases located in the neuromuscular synapsis can be corrected by the administration of cholinergic preparations such as edrophonium chloride (tensilon), prostigmine, pyridostigmine, etc.
Various laboratory tests, notably electromyography (EMG) and determination of motor nerve conduction velocity (MNCV), and tests for serum enzymes such as creatine phosphokinase (CPK), as well as histological, histochemical and electron microscopic studies of muscles explants are important tools for a more sophisticated differentiation of various neuromuscular disorders.

Neuromuscular diseases, i.e., disorders of the motor units, have to be distinguished from diseases resulting from alterations of supranuclear motor structures, i.e., motor nuclei and pathways above the peripheral reflex arch. Alterations of the corticospinal tract are usually characterized by spasticity. Best known in childhood is the spastic variant of cerebral palsy. However, not every case of spastic cerebral palsy is spastic from the very beginning. Cases of so-called spastic cerebral palsy may show considerable hypotonia for several months or even years, and in some instances, hypotonia persists. In the latter case, one speaks of atonic diplegia. It is distinguishable from motor unit disorders by the presence of the Babinski sign and hyperreflexia. Alterations in cerebellum and brain stem centers cause various types of supranuclear hypotonia. In these supranuclear hypotonias, a discrepancy between the decreased muscle tone and a normal or only slightly decreased muscle strength is noticed. Muscle weakness is not a common feature of this type of extrapyramidal, supranuclear hypotonia. Nevertheless, in some rare conditions, the hypotonia may be so severe that spontaneous volitional movements become impossible. Such conditions may be difficult to distinguish from lower motor neuron disease. One variant of severe supranuclear hypotonia—to be mentioned here—is the Prader-Willi syndrome, also called HHO or HHMO syndrome (H for hypometria, H for hypogonadism, H for hypotonia, and O for obesity). Peculiar anomalies of the teeth are found often in this syndrome, for which the dental surgeon may be consulted.

Prader and Willi, two Swiss pediatricians, together with the endocrinologist Labhart, described this syndrome for the first time in 1956. It is presumably hypothalamic in origin and shows two phases in its course. The first phase is seen in newborns and young infants. At this age, patients with the Prader-Willi syndrome present a state of severe depression of the central nervous system. The patients are completely motionless, flaccid and atonic. The face is flat; the mouth is triangular or tented. One speaks of the so-called fish or shark mouth. Tendon reflexes cannot be obtained in this phase of the Prader-Willi syndrome. Sucking and swallowing are not possible, and gavage feeding is necessary for a period varying from several weeks to several months. Boys with the Prader-Willi syndrome have a hypoplastic scrotum and hypoplastic penis and testicles are not descended. On
the basis of the latter, Prader-Willi syndrome can be suspected as early as the first phase. In girls it is difficult to distinguish the first phase of the Prader-Willi syndrome from other conditions of severe hypotonia.

After some months, infants with the Prader-Willi syndrome become more lively and responsive. They reach the second phase of this condition, which is characterized by delay of psychomotor development leading to a more or less marked mental subnormality. In addition, these children have a peculiar lack of emotional control. On one hand, they show friendliness and expressions of rapturous joy, which alternate with tantrums and episodes of excessive stubbornness on the other hand. Between two and three years, hyperphagia or polyphagia develops. Some patients with the Prader-Willi syndrome are plagued with persistent hunger which becomes a painful experience for them as soon as they stop eating. If regular food is not available, they eat the food of their dogs or cats, or even pilfer garbage cans. In rare instances, the patients are not able to experience the feeling of satiety. They continue to eat as long as food is in sight, yet do not actively attempt to procure it. As a consequence of incessant food intake, obesity of monstrous dimensions develops in some instances. Patients with the Prader-Willi syndrome will come to the attention of the dental surgeon because of the tendency to have decay of the deciduous teeth. In some instances, total decay of the crowns down to the roots is noticed. In other instances, the dental condition resembles amelogenesis imperfecta. The second dentition is usually not affected. The excessive obesity may lead to a Pickwickian syndrome with death due to hypercapnia and respiratory athetosis. Others develop diabetes and its complications. The prognosis of the Prader-Willi syndrome is guarded.

In the following, a few neuromuscular disorders will be discussed in which weakness of muscles innervated by the cranial motor nuclei prevails. Cranial nerve involvement is rare in the spinal muscular atrophies and the peripheral neuropathies, and if occurring, it occurs rather late or in the terminal stages of the disease process. It is occasionally seen in cases of very early (prenatally) beginning Werdnig-Hoffmann's disease, which progresses very rapidly, leading to bulbar palsy and death within weeks or months.

A congenital condition, the so-called Moebius syndrome, or nuclear aplasia, has to be mentioned at this point. This syndrome was first described by a German neurologist, Moebius (Leipzig), in 1888. The main symptoms are congenital facial diplegia and congenital sixth nerve palsy. Other symptoms are listed in Table 2. Moebius syndrome is found in both boys and girls. Familial occurrence is exceptional. It is said to be due to aplasia or degeneration in utero of the respective motor nuclei. However, in some in-
stances, a myopathy with dystrophic changes limited to some facial and ocular muscles has been described as well. The course of the Moebius syndrome is stationary (Table 2).

The most important and best known disorder of neuromuscular synapsis is myasthenia, which is not too rare a disease, and of which a gravis and a levior form have been distinguished. Kurland, some 15 years ago, estimated there were in the United States about 10,000 patients with myasthenia gravis. Myasthenia occurs in all ethnic groups and is more frequent in females than in males. Moreover, females develop myasthenia at an earlier age than males. Seventy-five percent of the affected females and only 25 percent of the affected males develop myasthenic symptoms before the age of 35 years.

The disease begins insidiously, although at times a sudden onset is observed. Early symptoms are facial diplegia, ptosis, double vision, dysphagia and dysphonia. Patients have difficulty pouting their lips. The weakness is less in the morning and grows worse as the day goes on. After a night's rest, the weakness may almost disappear, to re-occur during the course of the day. Weakness may also be noticed in the muscles of the neck and shoulder girdle, respiratory muscles and in many other muscles. Tendon reflexes may be active at the first attempt to elicit them, yet responses become weaker if the test is repeated a number of times. Repeated stimulation decreases muscle contraction.

The diagnosis, myasthenia, is confirmed by spectacular improvement of muscle function after intravenous administration of edrophonium chloride (tensilon). The improvement occurs within seconds after the injection and lasts for a few minutes only. A more sensitive test is the so-called curare test which, however, should only be used in patients who do not present a history of breathing difficulties. Myasthenic patients are very susceptible to curare and even an eightieth of the curarizing dose in normals produces a conspicuous effect. Neostigmine or tensilon for intravenous injection should be at at hand if the curare test is used. The average curarizing dose in normals is about 1.3 milligrams/kilogram body weight. If one-fifth of this dose does not produce a definite weakening effect, the patient does not have myasthenia or is in remission at the time of the examination.

The therapy of myasthenia consists of the oral administration of anticholinesterases. Neostigmine bromide (prostigmine) or pyridostigmine bromide (mestinon) are the drugs of choice. The latter is given in a four-times-higher dose than prostigmine. ACTH and corticosteroids have been claimed to have excellent effects, notably in children. Likewise, thymectomy has yielded good results, notably if the operation is done in earlier stages of the disease. Nevertheless, the prognosis of myasthenia gravis is guarded,
since exacerbation and therapeutic refractoriness may occur sooner or later.

Myotonic dystrophy or myotonia atrophica is a frequent disorder which is inherited as an autosomal dominant trait. The expressivity of the gene varies greatly. Some individuals are very mildly affected or present nothing more than laboratory evidence of the disease, such as EKG abnormalities or abnormal immunoglobulin levels. The disease can become apparent at any age, in childhood as well as in adulthood. It is characterized by myotonia, delayed relaxation of the contracted muscle. There is progressive muscular weakness which develops at a slow speed. Most patients are ambulatory for many, many years. The weakness initially is localized in the craniofacial muscles and the distal parts of the extremities. Facial diplegia causes a flat and sagging face which accounts, in combination with the hollow temples (atrophy of the temporal muscles), for the sad expression of these patients. The neck is thin and the head is bent forward. The speech is dysarthric and, in some instances, barely understandable. Dysphagia and nasal regurgitating occur and aspiration of food particles is a constant threat for the patients.

Myotonia atrophica is a multisystem disease with a number of other organs than the muscle being involved. Ocular findings include early polar cataracts, pupillotony, electoretinographic abnormalities and low intraocular pressure. The skull shows a thickened cranial vault. The myocardium may be involved and various electrocardiographic abnormalities are encountered. Cardiac failure and sudden cardiac arrests have been observed and can occur at any time during the course of the disease. Patients with myotonia atrophica represent poor risks for anesthesia. Various anesthetics, notably thiopentone, halothan and pethidine may induce respiratory arrests. Whenever a surgical procedure becomes necessary in such patients, it should be done under local anesthesia if possible, or if general anesthesia is necessary, under careful observation of the operative and postoperative period. Mental retardation is a frequent finding in myotonia atrophica, and many patients are defective with respect to their social responsibilities. They are inclined to defer the care of themselves to other persons or to social agencies. Various endocrine disorders, notably hypogonadism, occur in male and female patients. Immunoglobulins are low, as has been shown for the IgG immunoglobulins. This is due to an accelerated catabolic breakdown of these proteins. The smooth muscles may partake in the disease process and functional disturbances of upper and lower gastrointestinal tract, gall bladder and urinary excretory pathways have been described.

The muscle histology of myotonia atrophica resembles that of the muscular dystrophies, although ringbinden formation and central nuclei arranged in chains are often found in this condition. In the early phases of
the disease, atrophic changes prevail, which led some investigators to postulate that myotonia atrophica represents a neuropathy rather than a myopathy. However, blockage of the neuromuscular synapsis by curare does not alter the myotonia, which suggests that the basic pathology may still remain in the muscle.

In spite of the likeness of the histological picture to other muscular dystrophies, myotonia atrophica is to be considered as a particular disease entity to be classified under the myotonias rather than under the muscular dystrophies. There are several variants of muscular dystrophy which have to be considered as specific disease entities. Duchenne muscular dystrophy is the most frequent muscular dystrophy. Its incidence is about one in 6,000 live births. Duchenne muscular dystrophy belongs to the more severe and rapidly progressive myopathies. Facial structures are usually not involved in Duchenne muscular dystrophy although macroglossia due to pseudohypertrophy of the lingual musculature is often found. Duchenne muscular dystrophy is transmitted as an X-linked recessive trait. Boys are affected and females are heterozygous for the mutant gene. They are carriers or conductors of the trait, yet muscle weakness in females is very rare indeed. There are, however, various laboratory tests to detect carriers of the Duchenne muscular dystrophy gene, for example, elevated serum CPK, evidence of dystrophy in the muscle biopsy and abnormal in vitro protein synthesis of muscle ribosomes. One-third of the Duchenne muscular dystrophy cases are sporadic mutations. This has implications with respect to genetic counseling.

Muscular dystrophies with involvement of muscles innervated by cranial nerves are facioscapulohumeral muscular dystrophy, ocular and oculopharyngeal muscular dystrophy, and severe congenital muscular dystrophy.

The facioscapulohumeral dystrophy was first described by Landouzy and Dejerine at the end of the last century. It is transmitted in most instances as an autosomal dominant trait, which means that it can occur in several subsequent generations, that only one parent needs to carry the mutant gene in order to affect 50 percent of the offspring, and that boys and girls are affected with equal frequency. An autosomal recessive variant of facioscapulohumeral dystrophy occurs as well. More recently, a spinal muscular atrophy mimicking course and symptomatology of the facioscapulohumeral dystrophy has been described.

The disease can begin in infancy. Children affected with facioscapulohumeral dystrophy are not able to suck through a straw, to whistle or to blow out a burning match or candle. They have speech difficulties; notably the pronunciation of labial consonants is defective. The lips and perioral area are often somewhat thickened, which is perhaps due to pseudo-
hypertrophy of the orbicularis oris. They cannot close their eyes tightly, yet extraocular movements are intact. There is facial diplegia, but no external ophthalmoplegia. In some instances, facial muscles only are affected. In most cases, the disease progresses and leads to weakness of neck and shoulder girdle musculature. Later on, lordosis and weakness of pelvic girdle muscles appear. The progression of the facioscapulohumeral dystrophy is slow. We have seen patients who at the age of 45 years were still functioning as housewives satisfactorily, although considerable weakness in proximal extremities and truncal muscles with lordosis had developed. Life expectancy is usually not shortened.

The ocular and oculopharyngeal dystrophies represent other rare variants of muscular dystrophy. They may begin in earliest infancy. External ophthalmoplegia may be noticed in some cases. In other cases, ptosis alone exists, while again other cases show involvement of ocular and pharyngeal muscles. Histological studies of the eye muscles show dystrophic changes. We studied one case who had feeding difficulties and ophthalmoplegia since early infancy. He could not move his eyeballs at all. Moreover, he had a severe atrophy of his facial muscles with complete absence of any mimic expression. He could not close his mouth without the help of his hands. In order to chew, he had to push his fist against his lower jaw. His skeletal muscles were rather hypoplastic, and in adolescence he developed weakness in the extremities as well. Biopsy of the eye muscles confirmed the diagnosis of muscular dystrophy, yet serum CPK was normal.

Facial diplegia and more or less pronounced external ophthalmoplegia is also found in the severe congenital muscular dystrophy. Sucking and swallowing may be severely compromised and necessitate gavage feeding for a long time. Moreover, there is severe generalized muscular weakness and atrophy. Very few of these patients are ever able to walk. Some children with severe congenital muscular dystrophy show signs of arthrogryposis, the presence of which is indicative of severe muscle weakness and immobilization during intrauterine life. The muscle shows the typical alterations of muscular dystrophy with, at times, excessive endomysial fibrosis. The serum CPK is normal or slightly elevated. The condition is so severe that it is rarely compatible with a longer postnatal life. Almost half of the newborns with severe congenital muscular dystrophy die within the first year of life, and very few survive the first decade. Death is usually due to respiratory infection with consecutive atelectasis due to poor ventilation of the lungs. Severe congenital muscular dystrophy can affect more than one child in a sibship. It may be inherited as an autosomal recessive trait.

Improvement of laboratory techniques, notably in the realm of histological, chemical and electron microscopic examination of muscle explants
has led to the discovery of a sizable number of new myopathies. A selected list is given in Table 4.

In some of these myopathies, involvement of other organs may help in establishing a diagnosis. This is the case for the glycogenoses II, III and IV. In most other conditions, the clinical pictures are very much alike. The exact diagnosis is not possible without the sophisticated laboratory methods mentioned above. Referral of these cases to specialized centers is therefore mandatory, at least to establish the accurate diagnosis.

In this context, two conditions should be mentioned in which muscles innervated by cranial nerves are or may be affected, namely nemaline myopathy and the centronuclear myopathies. Both conditions are sometimes included under the heading congenital nonprogressive myopathies, although they are neither always congenital nor always nonprogressive.

Most patients with nemaline or rod myopathy are born as floppy infants with generalized muscular weakness, often involving facial muscles as well. The latter may lead initially to feeding difficulties which are overcome in later infancy. Motor development is somewhat slow, but they learn to walk and major physical handicaps are uncommon. Their body build is slender; skeletal musculature is poorly developed. The face is long, small and sagging. The mouth may be triangular with a tented upper lip. Tendon reflexes are diminished or absent. Sensory functions are intact.

The EMG suggests neuropathy. Serum CPK is usually normal. The diagnosis is made by the discovery of peculiar alterations of the Z-lines. The Z-lines fragment in the periphery of the muscle fibers and form coarse rectangular clumps which stain similarly to the Z-lines.

Nemaline myopathy is a hereditary disease transmitted as an autosomal dominant trait. The prognosis varies in accordance with the severity of the weakness, notably of the respiratory muscles. If the latter are severely compromised, early death as a consequence of pulmonary complications may occur.

Centronuclear myopathy, originally called myotubular myopathy, was first described in 1966. About 50 cases have been reported up to spring, 1973. In recent years, genetic heterogeneity of centronuclear myopathy was documented, suggesting that it consists of a group of diseases with similar histological alterations in the muscle fiber. The main pathology of the centronuclear myopathies consists of the centralization of the sarcolemmal nuclei, which are often arranged in long chains. The nuclei are surrounded by a halo of clear material which forms central vacuoles between the nuclei. These vacuoles consist, as documented by electron microscopic examination, of mitochondria, glycogen granules and debris of deteriorated myo-
fibrils. In some cases, only type I muscle fibers are affected, and in other cases both muscle fiber types are affected.

Most children affected with a centronuclear myopathy are born as floppy infants. At times, however, clinical manifestations are only noticed in childhood or even adulthood.

Histological evidence of centronuclear myopathy was found in some clinically unaffected relatives. Thus the disease can be clinically silent, at least for many years. The clinical symptomatology of centronuclear myopathy in general is very much the same as that of nemaline myopathy. Signs of facial diplegia are usually more pronounced and external ophthalmoplegia is a characteristic feature of roughly one-half of the cases. Weakness of mastication and swallowing and dysarthria are other typical, yet not constant, findings of the centronuclear myopathies. Some patients show an entirely different distribution of muscle weakness, which resembles more that of peroneal muscular atrophy. The intelligence of these patients is normal and sensory functions are intact. Serum CPK is normal or slightly elevated.

The prognosis depends on the severity of the clinical symptoms and also on the potential progression of the disease, which can be seen in a small number of cases.

Centronuclear myopathy can be transmitted as an autosomal dominant and also as an X-linked recessive trait. The occurrence of cases in siblings without any other affected relatives suggests autosomal recessive inheritance as well.

So far, neuropathies and spinal muscular atrophies have been strictly separated from the dystrophies and nondystrophic myopathies. The fact that in some centronuclear myopathies only type I fibers were affected suggested to W. K. Engel a possible neuropathic origin of the disease. Recent investigators suggest that there is some overlapping between lower motor neuron disease and muscular dystrophy. It was shown that some cases of scapuloperoneal and facioscapulohumeral dystrophy are indeed spinal muscular atrophies. It was furthermore found that the number of functional motor neurons in various muscular dystrophies is decreased. These findings suggest that a neuronal element plays a role and that the muscular dystrophies can no longer be considered as primary muscle diseases. It may be that we witness here a transient trend to mix up neuropathy and myopathy, which will disappear again as time passes. Yet it may also be that we are at the beginning of a new era, where an entirely new conceptualization of the neuromuscular diseases begins to emerge.

Summary

Neuromuscular disorders include diseases of the lower motor unit. These
disorders were briefly defined and a classification of them, including differential diagnostic criteria, presented. In the second part, a few neuromuscular disorders with involvement of muscles innervated by cranial nerves were briefly reviewed. Dentists should be familiar with the existence of these conditions, since they may be the first ones to be consulted for such cases.

Table 1

Differential Diagnostic Criteria of the Neuromuscular Disorders

<table>
<thead>
<tr>
<th>Disorder of</th>
<th>Serum CPK</th>
<th>EMG</th>
<th>MNCV</th>
<th>Muscle Biopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior horn cell</td>
<td>Initially normal,</td>
<td>Neuropathic*</td>
<td>Normal</td>
<td>Fascicular atrophy,</td>
</tr>
<tr>
<td></td>
<td>after 2-3 years</td>
<td></td>
<td></td>
<td>after longer duration, secondary myopathy</td>
</tr>
<tr>
<td></td>
<td>slightly elevated</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>in 1/3 to 1/2 of</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>the cases</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Motor nerve</td>
<td>Normal</td>
<td>Neuropathic*</td>
<td>Decreased</td>
<td>Fascicular atrophy</td>
</tr>
<tr>
<td>Neuromuscular</td>
<td>Normal</td>
<td>Mixed</td>
<td>Normal</td>
<td></td>
</tr>
<tr>
<td>synapsis</td>
<td>Normal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Muscle fiber</td>
<td>Varying from normal to</td>
<td>Myopathic</td>
<td>Normal</td>
<td>Various myopathic pictures</td>
</tr>
<tr>
<td></td>
<td>slightly elevated</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Myopathic features may appear in the later course of the disease as an expression of the secondary myopathy.
Table 2
Symptomatology of the Moebius Syndrome

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>61</td>
</tr>
<tr>
<td>Bilateral facial palsy</td>
<td>61</td>
</tr>
<tr>
<td>Sixth nerve palsy</td>
<td>45</td>
</tr>
<tr>
<td>External ophthalmoplegia</td>
<td>15</td>
</tr>
<tr>
<td>Ptosis</td>
<td>6</td>
</tr>
<tr>
<td>Weakness and atrophy of tongue (often unilateral)</td>
<td>18</td>
</tr>
<tr>
<td>Weakness of masticatory muscles</td>
<td>4</td>
</tr>
<tr>
<td>Clubfoot</td>
<td>19</td>
</tr>
<tr>
<td>Hand and arm malformations</td>
<td>13</td>
</tr>
<tr>
<td>Defect of musc. pectoralis</td>
<td>8</td>
</tr>
<tr>
<td>Ear deformities</td>
<td>8</td>
</tr>
<tr>
<td>Mental subnormality</td>
<td>6</td>
</tr>
</tbody>
</table>

From Becker, Humangenetik, 1968.

Table 3
Classification of the Muscular Dystrophies (MD)

X-linked forms
1. Duchenne MD (pseudohypertrophic)
2. Becker MD (slow motion copy of 1)

Autosomal recessive forms
3. Limb-girdle MD
4. Benign congenital MD (perhaps variant of 3)
5. Severe congenital MD
6. Facioscapulohumeral MD (rare variant)

Autosomal dominant forms
7. Facioscapulohumeral MD
8. Ocular MD
9. Oculopharyngeal MD
10. Ocular MD plus
11. Distal MD
12. Limb-girdle MD (rare variant)
Table 4
List of Selected Myopathies
Excluding Muscular Dystrophies and Myotonias

1. Central core disease
2. Nemaline myopathy
3. Centronuclear myopathies
4. Mitochondrial myopathies
5. Glycogenoses II, III, IV
6. Myophosphorylase deficiency
7. Lipid storage myopathy
8. Myopathy with nuclear inclusions
9. Red-ragged fiber myopathy
10. Familial myosclerosis
HOSPITAL DENTISTRY FOR THE HANDICAPPED CHILD

Ronald Johnson, D.D.S.

Advances in health care emphasize how the dental and medical professions can utilize the tools of scientific discovery with great success to further the ideal of better health for more people than ever before. The adoption of various insurance and other third-party payment programs will assure the payment of health care costs, bring about radical changes in hospital procedures and expand the role of dentistry in the total health picture.

In order to keep pace with progress in dentistry, the education of pre-doctoral and graduate students must emphasize the role of the modern hospital in good dental care. In the development of the hospital-oriented dentist, active responsible participation should be stressed. The tendency for dentists has been to avoid assuming the responsibility for hospitalized patients, and for physicians to avoid calling dentists in consultation because of the latter's reluctance to see patients in the hospital. This barrier can be eliminated to some extent by providing practical hospital experiences for dental students.

Great efforts have been directed toward oral care of the handicapped child and adolescent. Providing dental care for the physically and/or mentally handicapped is an important and sometimes difficult problem in the field of restorative dentistry, and is sometimes best met by hospitalization and full-mouth oral rehabilitation under general anesthesia. The modern hospital makes many services available, and through the efforts of the department of anesthesiology, provides a solution to the needs of the special or handicapped patient. It must be remembered that general anesthesia is not a panacea and should not be utilized in an attempt to solve every problem situation arising in the dental office. The doctor must not become...
complacent and should bear in mind the greater inherent risk to the patient with the use of general rather than local anesthesia. An attempt should be made to perform dental treatment for the handicapped patient in the dental office with the use of sedation and local anesthesia. Behavior modification techniques, premedication, nitrous oxide analgesia, restraining devices, mouth props, etc., are just a few of the many devices, techniques and procedures that are at our disposal. However, heavy and prolonged premedication can produce complicating postoperative problems. Although the use of local anesthetics can effectively block painful stimuli, the dentist must still cope with the problems of overriding anxiety and fear. Using nitrous oxide and oxygen as relative analgesia, the dentist has at his disposal an instrument which is safe, easily mastered and effective in its application to all phases of dental practice. The dentist who has experience and proficiency in administering nitrous oxide analgesia has an effective asset for the provision of complete dental care to the handicapped or exceptional child. If attempts using the above armamentarium are unsuccessful, general anesthesia should then be undertaken.

Many times after a patient has been treated under general anesthesia, he will cooperate to the extent of allowing treatment to be performed at the dental office in a routine way. When evaluating what type of patients may require a general anesthetic, the following categories are noted:

1. The physically and mentally handicapped (cardiac and hemophiliac, etc.);
2. The emotional or psychologically disturbed (autistic, psychotic, etc.);
3. The hyperactive and uncooperative child or adult;
4. The very young with rampant caries;
5. The patient who must travel great distances.

Dentistry for the patient being treated in the hospital under general anesthesia must begin with the principles of prevention. The need for a well-organized plaque control and nutritional counseling program has been shown to be especially critical for physically and mentally handicapped children. When general anesthesia is necessary to complete restorative procedures, it is important that a responsible individual have the oral hygiene of the handicapped child under control before the rehabilitation is attempted. Handicapped children usually are unable to care for their teeth adequately, and in most instances care becomes the responsibility of the parent, guardian or institutional attendant. In those situations where travel distance to the hospital or the severity of the condition prevents plaque control from being established before treatment, every effort should be made to work with the family during hospitalization and postoperatively to
establish an effective program for the child. Pit and fissure sealants, topical fluoride application and fluoride supplements should be considered in the total preventive program for the handicapped child.

When one is planning to do restorative dentistry, all of the procedures should be performed during the one visit to the hospital, even though several hours may be required. This eliminates the need of repeated admissions. The treatments that can be performed on these patients under general anesthesia are basically the same as those under a local anesthetic. These include silver amalgam, composites, stainless steel crowns, pulpotomies, extractions and impressions for space maintainers, castings, bridges, crowns, etc.

After it has been determined that treatment must be rendered under general anesthesia, an oral examination, radiographs and tentative treatment plan should be obtained before the patient is taken to the operating room. Even though these children frequently do not cooperate at this stage, an attempt should be made to determine the extent of the oral disease. In addition, the dentist should contact the patient’s physician to inform him of the planned procedure and to inquire about the child’s physical condition. After the patient has been anesthetized, a re-examination and evaluation should be made to determine additional findings and corrections.

A complete blood count should be performed during or prior to admission to determine if the patient’s hemoglobin, hematocrit and white blood cell count are abnormal. A routine analysis and chest film will also be required in most hospitals. If an abnormality appears, such as anemia or infection, the procedure should be canceled until the condition is corrected. When the patient is admitted to the hospital for the oral rehabilitation, he will be examined and evaluated by a staff physician. At this time, the necessary laboratory workup is ordered. Usually the patient is admitted the day before surgery, thus making it comparatively easy to prevent ingestion of solids or liquids and to administer the necessary premedication drugs.

No patient who has ten grams or less of hemoglobin should be anesthetized or he may develop a hypoxia which could result in depression of his myocardium. Any patient who has a white blood cell count of 10,000 or over should not be anesthetized until the cause of the leukocytosis is determined. The parents are urged to take precautions during the last few days prior to admission to protect the child from unnecessary exposure.

Although the administration of anesthesia is handled by the anesthesiologist, responsibility for it is still shared by the dentist. For this reason, he should be reasonably informed relative to the anesthesia procedures. It is not uncommon in certain patient problems for induction to be carried out with an explosive mixture and then a switch made to a nonexplosive mix-
ture for the operation. The dentist must make sure the anesthesiologist is aware that electrical equipment must be used in the procedure and the necessary precautions taken.

Five important steps of preparation make the course of the anesthetic smooth and permit the dentist to complete the restorative procedures with ease and continuity. These steps include the use of:

1. An endotracheal tube—which is the responsibility of the anesthesiologist,
2. Eye protection,
3. A pharyngeal pack,
4. A rubber dam,
5. A mouth prop.

When the endotracheal tube is passed through the nose, it is referred to as nasotracheal intubation; when passed through the mouth, orotracheal intubation. It is usually best to have it understood in advance that nasotracheal intubation is to be employed if at all possible.

Strict attention must be given to the protection of the patient's eyes. Ophthalmic ointment, nonadhesive tape, a 2x4-inch piece of moistened gauze or a sheet of rubber dam placed over the eyes and taped to the face are methods used to protect the eyes during the procedure. If the eyelids are not kept closed, there is danger of fragments of tooth, bone or restorative material getting into the eyes and causing a laceration of the cornea.

A clean technique is used for most oral rehabilitation procedures, so a final thorough scrub is indicated. Rubber gloves are usually not worn. However, where many stainless steel crowns are to be cemented, gloves may be used in order to keep the sticky cement off the operator's fingers. Before the patient is draped, a doughnut support is placed under the head for stabilization and towels are folded under the shoulders to improve the position of the patient for oral access. A large sheet is extended over the patient from the foot to the chin. Two towels are placed obliquely across the patient's chest on the right and left sides below the chin.

After draping is accomplished, the endotracheal tube is positioned over the patient's forehead with foam rubber pads or folded towels. Adhesive tape is wrapped around the tube and secured to the patient's face in order to maintain its position and depth into the trachea. The endotracheal tube extends over the right eye and the right side of the forehead. Whether the patient is draped (head and/or body) seems to vary in different hospitals. Some dental teams feel it gives added protection to the patient's body, face and hair, while preventing unnecessary cross-contaminations.

Before or during the induction procedure, the anesthesiologist will us-
ually tape a stethoscope to the patient's chest and place a blood pressure cuff on the arm. Intravenous fluids will usually be started by the anesthesiologist, but he may request the dentist to start this procedure. Usually five or ten percent dextrose and water will be started intravenously in an arm or foot vein.

In restorative dentistry, one is concerned with a pharyngeal or throat pack which is used in conjunction with endotracheal intubation. This pack prevents tooth fragments, restorative material, calculus, mucus or blood from passing into the pharynx and perhaps eventually into the trachea. The packing of the mouth and throat is the responsibility of the dentist and not of the anesthesiologist. The pharyngeal pack should have an attached string or tape long enough to extrude from the oral cavity. This string is used as a reminder that a pack is present in the throat. After the oral pharyngeal area is carefully suctioned, a moist 4x4 or 18-inch vaginal gauze is introduced with tissue forceps or hemostats into the throat in front of the endotracheal tube and behind the posterior pillars. If placed properly, it will not interfere with the dentist's field of operation. Usually the pack does not require changing during a procedure. However, if it does become saturated with mucus or blood, it may be replaced. The use of the endotracheal tube and the rubber dam does not preclude the use of the throat pack. It must serve as a seal and protective barrier.

In recent years, few articles in the dental literature have placed emphasis on the effective positioning of equipment and members of the dental team in the hospital operating room. According to Davis et al., 1968, the design and positioning of equipment may have to meet specific hospital standards, and in most instances, the equipment must be mobile and versatile enough to permit other procedures to be performed in the same area. However, every effort should be made to adapt the hospital operating room to meet the needs of the rehabilitation team to increase efficiency and save critically needed time and energy. Dental equipment must be positioned to simulate the dental office operatory. Sit-down four- or six-handed dentistry utilizing one or two assistants has proven to be the most effective means of accomplishing an oral rehabilitation.

Whenever possible, the dental team should be seated throughout the procedure on well-padded stools with adequate back support. Some hospitals permit the use of a contoured dental chair instead of the traditional operating room table. This chair may be permanently located in the room or easily moved into position when necessary. However, in most instances, the dentist must utilize the available operating room table and adapt his techniques to fit the available facilities.

A major problem in positioning the dental team is the discomfort experi-
enced by the first assistant while leaning over the operating table in order to achieve adequate visibility of the patient's oral cavity. In many instances, the problem is created by the width of the operating table. One solution is to place the patient obliquely on the table with his head closer to the first assistant. If the operating room is small, the table may be obliquely positioned to allow additional mobility for the assistants. Another solution is to alter the head of the operating table by attaching a removable head and back board extension which is secured by ordinary thumb screws. These table modifications allow the anesthesiologist ready access to the controls and comfortable seating for the assistant and operator. In most hospitals, operating rooms are larger than dental operatories, so, fixed cabinetry against the walls is a definite disadvantage. Utilization of preset trays has been found to be ideal for instrument placement and for maintenance of a clean organized work area. A Mayo stand, which extends over the chest of the reclined patient, may be used to hold material and instruments for prepping the patient or may be utilized as the medication and instrument tray throughout the procedure. When used for prepping, the following items will be found on the tray: throat packs, material for eye protection, and body and head drapes. A mobile operative unit positioned in front of the first assistant should be equipped with high and low speed handpieces, an enclosed central evacuating system and an adequate work area in which to place preset trays. Because of their mobility and illumination, one or more high intensity ceiling-mounted lights are recommended for large operating rooms.

The dental team may vary from a two-person approach with one operator and assistant to a three-person approach with one, two and sometimes three trained operators. During extremely long procedures, the three-person approach has the advantage of allowing the rotation of the team positions, thus reducing fatigue that might affect optimal results. For the two-person dental team, an additional circulating assistant may be utilized to mix materials, prepare instruments and aid other aspects of the treatment, thus simulating the three-person team approach. In either method, the first assistant's duties are confined to tissue retraction, maintenance of a clear operating field, instrument transfer and other functions that directly aid the operator. The second assistant manages those procedures which are outside the working area of the oral cavity, such as mixing restorative materials and medications, preparing stainless steel crowns, etc., and is positioned opposite the first assistant between the mobile cabinet and the operative unit.

Most operators using sit-down four- or six-handed technique prefer to sit in the area between 10 and 12 o'clock (patient's head related to the hour
positions on the face of a clock). Thus, the anesthesiologist may be seated at 9 o’clock and the first assistant at 3 o’clock. It is felt by some dentists who work in a hospital setting that positioning of the operating team should be determined by the dentist for his convenience. However, in looking for maximum efficiency, all members of the operating team should be considered in the selection of proper positioning.

A mobile cabinet with supply drawers may be converted to a completely functional operative unit by mounting on it, in position for ease of access, the following items: high and low speed air-driven handpieces, triplex syringe and high velocity suction. However, there are various types of manufactured equipment on the market which have extensive mobility and flexibility and will enable the dental team to establish any variation of a two- or three-person approach while working in a sit-down four- or six-handed technique. Thus, careful planning of the procedure, selection of equipment and effective positioning of the dental team must be considered in order to obtain maximum efficiency, speed and better dental care for the patient.

It is hoped that some aspect of this presentation will be of aid to the dentist using the hospital to render care for his patients. Perhaps he will take a moment of his time to evaluate the operating room and the organization of his dental team.
ANESTHESIA AND ANALGESIA:

ADJUNCTS IN THE DENTAL MANAGEMENT OF THE HANDICAPPED CHILD

Harold Turner, D.D.S.

A review of the medical and dental literature over the past twenty years readily reveals that multitudes of papers have been written on the assign-ment of anesthesia and analgesia for the dental management of the handi-capped child.

It would be presumptuous of me, in addressing the participants of this symposium, to espouse the use of a personally favored, and perhaps mas-tered, technique or pharmaceutical aid as the only acceptable methodology, to the derogation of the many others available.

However, it would be equally derelict of me if I were not to present my attitudes and prejudices acquired in actively delivering dental care to the handicapped child for twenty years.

My discussion today is oriented toward the use of the forementioned ad- juncts within a dental office environment or in an outpatient ambulatory service.

At the outset I must state that my basic philosophy decries the label "handicapped child" and its concomitant stereotype suggesting the constant need for heroic measures in the approach to dental care. For some reason not apparent to me, there still prevails an aura of mystery and mystique in the delivery of care, a feeling of apprehension and uneasiness for both the professional and the parent.

When we, as the caretakers of children within a professional expertise, embark on uncharted or perhaps poorly or incompletely charted waters,
then it is incumbent upon us to develop a methodology to render optimal services based on knowledge and specific information, for ultimate understanding of the problems presented. There are no magic potions, nor is there any mystery in the care of the so-called handicapped child.

We know that many children have special needs and/or requirements. We must accept each child as he seeks care on an individual basis, and if he should present a condition or situation not consistent with society's definition of normal, then we, the professional caretakers, must mobilize all of our knowledge, experience, information, aids, adjuncts and techniques to tailor treatment for this person to assure optimal oral and dental health. The delivery of care must be fully consistent with the needs of the child and not compromised in any manner.

Anesthesia and analgesia as related to ambulatory dental treatment are considered within the context of psychosedation, and in their full scope include all adjuncts, aids, drugs and techniques that effect treatment in a conscious, semi-conscious or unconscious state.

Anesthesia and analgesia are effective and efficient adjuncts in the dental treatment of the child with special needs, if basic requirements are set and adhered to.

A complete medical history must be taken, reviewed, properly evaluated and assessed prior to the institution of any treatment. History taking starts with the initial telephone call, at which time the following information is obtained:

**History**

1. Name of patient, including nickname.
2. Address of patient.
3. Age, including birthdate.
4. Sex.
5. Grade in school.
6. Purpose of call (e.g., pain, neglected treatment, unmanageable).
7. Parents' objective view of handicap, and general physical and dental conditions; also, time of day when patient is most restless. Is patient taking any medications?
8. Name and address of person referring patient.
9. Name, address and phone number of previous dentist, if not referrer.
10. Name and address of pediatrician or physician.

Referrer, physician and dentist are called to gather preliminary information to enable office personnel to properly prepare themselves for the patient.

At the initial visit, in an atmosphere that is calm, and in a manner generating honest concern and interest, the parent(s) (and child) are interviewed and a fully detailed history is taken.
Ask specifically about the impairment:
The diagnosis as the parent knows it;
When diagnosed, by whom, and where treatment has been sought and instituted;
Effect of child’s behavior pattern:
   - Attitude toward doctors, hospitals,
   - Parents’ attitude toward same,
   - Attitudes toward family,
   - Parents’ attitude toward child.

Cooperation is most important and parents must know reasons for treatment (are they ashamed, overprotective, aware of the situation?).

Toilet training.

Feeding.

Education: intelligence level, grade in public school (or private), general problems especially noted in communication and in function, e.g., muscular coordination and hyperactivity.

Speech.

Vision.

Hearing.

Any known dental or oral facial deformities.

Any known pharyngeal deformities.

Oral habits.

General habits.

Diet.

Oral hygiene.

Ask specifically about possible medical complications:

Does patient have or have you ever been told he has had:
   - Heart trouble (or murmur),
   - Diabetes (sugar in urine),
   - Kidney trouble,
   - Asthma (or allergy),
   - Breathing difficulties (tuberculosis),
   - Liver disease (yellow jaundice),
   - Epilepsy (seizures),
   - Fainting,
   - Bleeding problems (does a cut heal? bruise easily?),
   - Does he take iron,
   - High or low blood pressure,
   - Any skin disease,
   - Arthritis.

Ask specifically about drugs:

Is patient being treated by a physician now, if so, what drugs is he taking?

Has he ever had any drugs for:
   - infection (any reaction to penicillin, tetracyclines, sulfa?);
sedation (are they effective?).

Has he ever taken, and what was the effect, if any, of:
- barbiturates,
- tranquilizers, especially phenothiazines (potentiate action of sedatives),
- Rawoulphia drugs,
- steroids,
- amphetamines,
- anticoagulants,
- diuril,
- antiasthmatics,
- local anesthetic,
- insulin,
- belladonna derivatives, etc.

Ask "Is there anything else you wish to tell me?"

E.g., patient may now have a bruised area or sensitive area which you may accidently touch, or be adverse to certain words, phrases or colors.

After the history has been taken, the child (if possible) is ushered into the operatory for clinical oral and radiographic examination. It is at this time that the child should be allowed to engage in verbal catharsis.

It is at this time that the operator will be given the clues for assignment of the best methodology for management. We try to utilize the simplest approach consistent with the child's needs, but we are constantly aware and alert to possible alternative approaches when it becomes evident that our initial methodology will not allow us to treat successfully.

The most important fact to bear in mind at all times is that we are there to deliver dental care, rather than to set a goal of proving or disproving the effective use of adjuncts—if one technique proves ineffective in a maximum of three attempts, then an alternative should be instituted immediately. Dental disrepair and oral disease are functions of time, and control of disease will not be effected while individual idiosyncrasies (operator or child) are satisfied.

To reiterate, the behavior of the child in the chair will ultimately determine the best method of approach. It will be found that after gathering the necessary information by means of a thorough history and straightforward attempt at establishing rapport with the child, most children with handicaps or special needs can be treated effectively and efficiently within the normal routine of the dental office.

If after a sincere effort has been made, or it is sufficiently evident from gross inspection and personal experience that pharmacy should be utilized, then the proper drugs and technique consistent with the health of the child are delineated for use. In many instances what seems to be a radical approach may be the most conservative.
The operating environment and personnel must be fully prepared for treatment with adjunctive medication. The staff attending this patient should make themselves fully aware of the drugs and techniques that will be used and should be comfortably knowledgeable about the known physiological actions of the drugs being used, and especially aware of the known contraindications and adverse reactions. The staff should be and feel fully confident in situations of emergency, have all the necessary drugs and equipment readily available, and be knowledgeable in their use to counteract any and all adverse reactions with speed and efficiency. Always remember that the use of any agents for anesthesia and analgesia imposes an insult to the physiologic integrity of the patient, and only agents whose actions are speedily reversible should be used.

In my experience, it has been found to be sound policy to select a few drugs and techniques and learn them well, rather than to encourage the use of a plethora of aids simultaneously, thereby superimposing unknown, or rather unanticipated, reactions. Each additive requires closer total monitoring.

If analgesia alone is used, the office should have all equipment and supplies necessary for the maintenance of a patent airway and for control and/or support of respiration and circulation. A good supply and delivery system of oxygen must be at hand, as must an efficient suction apparatus. If, however, the operator wishes to institute more sophisticated total anesthetic techniques, then the office environment must be no less prepared than the hospital environment, and the child treated in no different manner than he would be in a hospital. More will be said later in this discussion regarding patient preparation, anesthetic techniques and recovery precautions for the office general anesthesia.

Analgesia is regarded by many proponents within different definitions. Semantics has played a great role in the teaching and use of analgesia. To some it bears the connotation of absolute freedom from pain, while to others it bears the requirement of inducing true sedation, and to still another group it serves only as a placebo either for the patient or the operator, or perhaps for both.

Analgesia within the context of my personal experiences and prejudices means psychosedation in its fullest meaning. Its delivery connotes the state of the patient as conscious or semiconscious, and it is expressed as the introduction of pharmaceutical aids in all their modalities. When I first became acquainted with analgesia it was then referred to as premedication, comedication and ultimately balanced medication. Its usual and accepted routes of administration were primarily P.O. (orally), S.Q. (subcutaneously), or I.M. (intramuscularly), and rarely P.R. (rectally) or I.V. (intra-
venously). Inhalation technique did not rear its head at that time because of the disrepute of nitrous oxide use in dentistry.

It has been refreshing to observe the advances in the philosophy and approach taken by our profession. Twenty years ago, you might have been considered a maverick for using drugs beyond local anesthesia for dental management, and then it was done only by the oral route; those who dared explore the other routes of administration were considered radicals and viewed with suspicion. It was totally inconsistent; a dentist adept in the use of a needle and syringe for the administration of local anesthetics in the oral cavity was considered less competent to learn the pharmacology of other drugs and how to use the same or a similar syringe subcutaneously, intramuscularly or intravenously to enable him to deliver total care for a patient who could not or would not have it done any other way.

For many years, intravenous techniques for sedation and anesthesia were considered only within the province of the oral surgeon, and because of the great experience with I.V. techniques in oral surgery, the profession itself is advancing its educational base to include I.V. sedation for all types of dentistry.

In evaluating the various modes of administration of analgesia, my own experiences have prejudiced me against the use of any modality wherein I do not have a measure of control over the action or interaction of drugs. Oral and/or intramuscular administration of drugs to produce required sedation and pain control has produced less than pleasant experiences. I have found that it requires more than one visit to establish a good baseline dosage. The actual time lag for drug action is most variable. Dosage determination by any and all methods proposed is at best most inaccurate, and individual daily idiosyncrasies (patient and operator) made dosage application even more variable. In substance, I could not accomplish my treatment goals in a fast efficient manner without compromise. Many visits were required to accomplish what could have been done in few visits. Each visit with oral or I.M. medication was insufficiently productive for the insult imposed on the physiology of the child; at desired depths of sedation the child was actually in chemanesia requiring supportive treatment. There were frightful postoperative periods. I reiterate that the forementioned is my personal prejudice.

Over the years, and after many thousands of trials with various drugs and techniques, I have assigned my priorities of administration when (and only when) consistent with the special needs of the child.

If warranted, I will utilize oral, intramuscular, subcutaneous or rectal administration, whichever is best for the situation at hand for induction, knowing that sedation will be maintained by inhalation or intravenous tech-
niques. Where feasible I prefer the rectal route to all the induction or primary sedative approaches. The time of onset is usually quicker and somewhat more predictable than oral or intramuscular, and although total absorption and metabolism is less predictable, the patient is being supported or maintained by other, more reliable, predictable and reversible methods during the time of the initial drug uptake. My preference for rectal administration is either pentothal sodium or brevital.

Intramuscular administrations are primarily reserved for situations wherein I desire a slow release and prolonged action of a drug concomitant with an intravenous or inhalation administration.

When using the intravenous modality, I try to restrict my armamentarium to two or three drugs, consistent with the physiologic indications and contraindications of the patient. Two intravenous infusion sets are set up, one with dextrose and water to be used for hydration and a patent pathway, and the other with a dilute solution of .01 percent or .02 percent pentothal sodium; all other drugs are introduced as required by bolus injection via the tubing. All drugs are submitted to an initial test dose and observed reaction (with careful monitoring of vital signs), and are then titrated as indicated until desired sedation is reached. The patient is responsive to stimulation at all times and is never brought to a depth where reflexes are obtunded. Local anesthetics are supplemented when indicated. The drugs of choice at this time are (diazepam) valium alone or in combination with pentothal sodium drip and/or nitrous oxide and oxygen.

Intravenous sedation is a sound efficacious technique, providing the patient will tolerate the intravenous injection and there is an accessible vein with sufficient integrity.

Nitrous oxide/oxygen administration as practiced today is, I feel, the most efficient, effective and safest modality for psychosedation. It should be emphasized that the use of either intravenous or nitrous oxide/oxygen techniques requires at least a minimal level of communication between operator and patient, and an airway that can be kept patent with minimal effort, and that although the levels attained for sedation may be conscious or semiconscious, these techniques are not meant to substitute for full general anesthetic techniques when they are indicated or required.

Trieger and Carr in their excellent review of nitrous oxide sedation set forth the most efficient methods for the use of nitrous oxide/oxygen psychosedation. Psychosedation with nitrous oxide and oxygen is accomplished by the effects created in the central nervous system by the inhalation of controlled amounts of nitrous oxide and oxygen, and by their potent effect as a placebo. Nitrous oxide/oxygen and/or intravenous psychosedation may be used in dentistry for a variety of procedures, i.e., radiographic surveys, pro-
phylaxis, impression taking, reducing gagging and providing a more cooperative patient. Operative dentistry, crown and bridge and periodontics can be done, though it is important to use local anesthesia as an adjunct where the operation is anticipated to be a painful one. Extractions and periodontal surgery would generally require adjunctive local anesthesia.

In none of the other modalities do we have an agent that produces onset of signs within 45 to 50 seconds, nor can the level of sedation be controlled and altered as rapidly as with nitrous oxide/oxygen sedation.

It is worthwhile noting that in none of the other techniques do we have a return to normal as rapidly (120 to 180 seconds) as at the conclusion of the nitrous oxide/oxygen session.

The record of safety experienced in the use of nitrous oxide/oxygen for conscious patients is remarkable. No serious complications have been reported in the performance of many such procedures in the past few years. Important consideration must be given to the concentration of oxygen in this technique. Psychosedation can be achieved and adequate oxygenation assured if the concentration of oxygen is never lower than 25 percent or 2.5 liters per minute flow. Jaffe, in a recent searching paper, reviewed the literature and concluded that “nitrous oxide used properly is still the least toxic of all anesthetic agents.”

As previously stated, the initial absorption of nitrous oxide is extremely rapid. Approximately 1,000 ml. at a normal respiratory volume is taken up within the first minute. At the end of ten minutes, it drops to 350 ml., drops further to 200 ml. in thirty minutes, and to only 100 ml. in one hundred minutes. Studies by Eger show that breathing 100 percent nitrous oxide will require one minute to reach 80 percent alveolar concentration! At one percent nitrous oxide, it takes four to five minutes to reach 80 percent inspired concentration.

It is readily demonstrable that it generally takes approximately 40 seconds of breathing nitrous oxide for patients to experience sensory changes such as tingling, feeling “high,” feeling warmth and other changes in perception. (When treating a patient with a handicap, the operator should make a determined effort to explain the reactions to the drugs used and to explain that the effects are desired. Constant communication and suggestion are necessary during induction and maintenance.)

Patient variability in response to nitrous oxide is very great. Each patient must provide his own titration. There can be no arbitrary or predetermined levels, since we are dealing with individuals who vary tremendously in their response to the percentage of nitrous oxide.

The patient is started on 100 percent oxygen, and nitrous oxide is introduced one liter at a time, while the oxygen level is reduced by the same
amount, until the desired level of sedation is attained. Generally 50 percent nitrous oxide or less is required; however, never go below 25 percent or 2.5 liters of oxygen.

The elimination of nitrous oxide follows a reverse pattern of uptake. A large volume is removed initially. This high initial outpouring of nitrous oxide may produce hypoxia (diffusion anoxia) on recovery by displacing oxygen. Though this is not usually found with nitrous oxide/oxygen sedation, following general anesthesia there is a tendency toward hypoventilation on recovery, as carbon dioxide is blown off and the respiratory rate falls. This diffusion hypoxia is most apparent during the first five to ten minutes in recovery. Oxygen administration at the end of each procedure is advisable.

In the total consideration of dental management of the child with a handicap, we are all realistic and fully aware that many of these children cannot be managed except while in an unconscious state with the administration of a general anesthetic.

The question arises every so often whether administration of a general anesthetic should be accomplished in the dental office or in the hospital. I can stand here smugly and say it should only be done in the hospital, because I practice in an area where I have access to operating room privileges at nine hospitals; but what about the practitioner in a geographic area where hospital facilities are not available? Or let us assume he is in an area where there is physical but not professional access to a hospital. These are points in fact and realistically affect the delivery of dental care for the child with special needs.

There is no debate regarding the site of treatment for children with medical problems requiring a hospital environment when stress of any sort is imposed on or infringes upon the physiologic integrity of a less-than-medically-sound patient. When we discuss general anesthesia in a dental office it is acknowledged that we refer to a good-risk patient without undue medical problems.

Patient preparation and office requirements are more stringent when general anesthesia is used to render the patient unconscious for prolonged periods of time.

The patient must have a good physical evaluation, and all indicated laboratory procedures (blood and urine) should be done prior to treatment (preferably within 24 hours). At the appointed time for treatment, the parents and child are questioned regarding food or liquid intake (instructions are given prior to the appointment that nothing is to be taken by mouth from midnight of the night before the appointment), the child's temperature is taken, and an immediate preoperative heart and lung exam is done. If all systems are go, then the child is taken into the operatory. If
he is very young mentally, and if the situation permits, he is induced either while sitting in his parent's lap or with the parent present—in whatever manner is consistent with as atraumatic an experience as possible.

Induction techniques range from inhalation utilizing nitrous oxide/oxygen, to halothane/oxygen, I.V. pentothal, rectal pentothal sodium, or brevital. Each situation is considered at the time, and the most appropriate method is used.

After induction, an intravenous infusion set is always set in operation for hydration and as a patent pathway for medications. Anesthetic maintenance is accomplished by either inhalation or intravenous agents with oxygen insufflated. During the total treatment time, the patient's vital signs (blood pressure, pulse and respiration) and an electrocardioscope are continuously monitored.

The patient is properly positioned, mouth propped open, a suitable posterior pharyngeal curtain is placed and the treatment is instituted.

It should be noted at this time that the staff attending this patient is concerned only with this patient and set for efficient completion of all indicated procedures to attain optimal oral and dental health. If the patient is physiologically stable and all personnel work in coordination, there are no restrictions to any dental procedures and no reasons for not completing all the necessary dentistry in a single visit.

A few remarks should be made concerning the anesthetic techniques themselves. Over a period of five years we randomly assigned techniques of anesthesia maintenance ranging from straight intravenous without oxygen insufflation, to insufflation of halothane and oxygen, nitrous oxide/oxygen with or without a nasal catheter, and endotracheal intubation. In my experience, the most efficient and least hazardous method is utilizing endotracheal intubation. We are all fully aware that there may be complications with endotracheal intubation and a reported high incidence of postoperative laryngitis and pharyngitis. However, experience has dictated that there is as high if not higher incidence of pharyngitis and atelectasis postoperatively with insufflation techniques, as well as possible complications of aspiration during the induction and during emergence.

When general anesthesia is used in the office, it is of prime importance that good recovery facilities including oxygen, suction, emergency drugs and expert personnel be available. The patient is allowed to leave the office after examination (especially pulmonary) and observation that all vital signs are stable and all reflexes have fully returned. The patient is called in the evening and on the following morning for follow-up.

In my own office, the operating team consisted of an anesthesiologist, a
dentist (fully trained in general anesthesia), an assistant at the chair and one circulating.

General anesthesia is an efficient, efficacious method for dental management of the handicapped child when other management methods would prove to be less than desirable.

In summary, delivery of dental care of the highest quality can be effected for all children with special needs, or so-called "handicapped children," with the judicious use of analgesia and anesthesia.

References

A NEW DIRECTION IN THE TRAINING OF PROFESSIONALS FOR DELIVERY OF SERVICES TO THE HANDICAPPED

Rudy Hormuth

Despite the almost universal focus of the news media these days on aspects of the Watergate affair, I am certain that some bits and pieces of information relating to a proposed major reorganization of the health components of the federal establishment have filtered through to most of you. The health services of the Department of Health, Education and Welfare are currently undergoing some major changes.

For the purpose of increasing the efficiency and effectiveness of the Department's health programs and in order to facilitate the development of sound policy in the area of the Department's responsibility, a number of agencies are being restructured. This restructuring includes the grouping of all health programs in the Department into five major organizational units—the Center for Disease Control, the Health Resources Administration, the National Institutes of Health, the Food and Drug Administration and the Health Services Administration. This new structure is designed to facilitate previously announced administration decisions to move in the direction of getting the federal establishment out of the business of some direct service operations and transferring more of this responsibility to where the "action" is, via revenue sharing, decentralization, and greater state and local responsibility. In some instances, a reduction in force of the federal establishment, as well as a refocus or termination of established programs that have either
achieved their purposes or are no longer relevant in these times, is contemplated.

These contemplated changes may ultimately have a major impact on basic manpower training as previously supported by the Department. A ripple effect on the curriculum of your own professional schools is likely via an intensified re-evaluation and reappraisal of goals, objectives and methods. Likewise in this new scheme of things, manpower training as such may not ultimately be defined as an appropriate federal priority activity. Administrative mechanisms closer to the service delivery system may well have to assume increased responsibility for basic manpower training. In that event we can anticipate some major changes in the goals and objectives of most professional training.

In preparing to meet some of these projected changes, reassessments, re-evaluations and consequent need for the restructuring of some training goals and objectives, we should keep in mind some of the recently-evolved models of service delivery which undoubtedly will have an influence as change agents in terms of the kinds and levels of personnel that will be demanded by the service delivery system. The impact of such concepts as comprehensive care, health maintenance organizations, the use of aides, para- and sub-professionals on a revamped manpower training program can be extreme.

In all of this there is no doubt that the concept of interdisciplinary training will come to play an increasingly major role at all levels of most professional training. While this is still a rather ill-defined curriculum concept, the term interdisciplinary training has emerged in the literature within the past few years primarily in educational programs in child development and on a broader scale in relation to the university-affiliated programs. The concept stands high in the priority listings of both the Department of Health, Education and Welfare and most universities.

Webster simply defines interdisciplinary as something which involves two or more academic disciplines. In order to clarify the slippery semantics of interdisciplinary training, one almost needs to construct a composite definition and include concepts from such words as train, trainer, educate, educator, discipline, inter, multi, professional, etc.

John H. Meier of the JFK Child Development Center at the University of Colorado Medical Center in Denver has attempted to construct such a conglomerate definition. He defines an interdisciplinary or interprofessional educational program as an educational program staffed by a group of persons skilled and informed about different aspects of a general field of endeavor who pool their talents and knowledge for the reciprocal benefit of
students who are interested in learning their skills and information as a means of livelihood.

This description of what interdisciplinary training is all about requires each academic profession to say who and what the skilled persons and talents are that should be pooled for the benefit of students. What can students in dentistry and pedodontics get from other disciplines, and what can your academic profession provide to students in other disciplines which will ultimately help this newly-emerging manpower be more responsive and more capable of efficiently and effectively dispensing services within the framework of the newer systems of service delivery?

A clearer understanding of what may or may not be involved in interdisciplinary training can be arrived at by exposing some of the roots of the concept and by tracing some of its early developments.

The major essential ingredient which was required to set this concept into motion was a problem, related to the delivery of care, which was so massive and extensive that it could not effectively be dealt with through any type of solo practice, nor could it be dealt with by a single academic profession or by any one agency involved in the service delivery system.

The problem which presented itself and which provided the basic impetus for the development of interdisciplinary training as a formal concept was that of mental retardation. It was the problem of mental retardation, however, as it began to be viewed after World War II. Admittedly as a condition, mental retardation has always been with us. Bone fragments from primitive man clearly establish the presence of microcephaly and hydrocephaly. History documents the presence of idiots and monstrosities at times when it was fashionable to retain such individuals among the household slaves. In those early times, however, the condition of mental retardation was not really a problem. Most inadequate individuals did not survive. The few that did were protected so that they could serve to amuse the more fortunate. It was not until after the beginnings of urban development, of industrialization and the assemblage of numbers of inadequate individuals in asylums and workhouses, that society began to become aware of mental retardation as a problem. The efforts of such early leaders as Pinel, Stard and later Seguin to "cure" this group or make them normal succeeded chiefly in identifying vast numbers of these inadequate individuals, and ultimately resulted in this country in the development of institutions far from population centers where these human failures could hopefully be hidden and forgotten.

Following World War II there was the emergence of a new climate in which the dignity of the human being was greatly enhanced. We had fought for and sustained basic principles relating to the worth and rights of human
beings. This country found itself in the midst of a baby boom. New breakthroughs in the care of infants were applied in our health care system. We succeeded in salvaging an unprecedented number of inadequate damaged babies. Mental retardation as a problem of enormous dimensions became visible. Parents who had mentally retarded children likewise became visible to each other. They began to compare notes and found that other parents had experienced the same exclusion from services as they had. Too many professionals had offered only one alternative—institutionalization—to too many parents. As a self-protective measure local groups of parents began to organize. As organized groups they began to demand services. This movement culminated in the formation of the National Association for Retarded Children in 1950. This national body then began to act as an advocate for all parents of handicapped children. They wanted inclusion of their children in our service system. They wanted alternatives to residential warehousing of their children.

The basic foundation for an interdisciplinary approach to the problem of mental retardation and the consequent need for interdisciplinary training of professionals to fulfill this objective was laid down in 1955. In that year four major events occurred:

1. The five-year-old NARC, now a strong national leadership group, developed and presented to the federal government through Congressman John E. Fogarty and the House Appropriations Committee a proposed program of national action in behalf of the mentally retarded. The document detailed how aspects of the array of available human services could be applied to meet the needs of the retarded and how such application might alleviate a good portion of the problem. Congressman Fogarty in the appropriation hearings of that year used the document to question the federal agencies that came before his committee as to what specifically they were doing in behalf of the retarded and alerted them to the fact that he would repeat these same questions in the budget hearings for the following year. A most important aspect of this NARC document and Congressman Fogarty's apparent concurrence with its approach was the fact that it did not relate needs to a single discipline or to a single constituent agency of H.E.W. Rather, it highlighted specific contributions to a total comprehensive care program that an array of professions and programs could and should be making.

2. The Department of Health, Education and Welfare, in response to demands of groups such as NARC and in a real effort to respond to unmet needs, in 1955 set up a Secretary's Committee on Mental Retardation. Here again, in what was something of a departure from the usual procedure of assigning responsibility for the development of policy, Departmental posi-
tion and response to a particular problem to a “lead” agency, the Department utilized a committee of representatives from various constituent agencies, responsible to the Secretary, to evolve position, policy and response. The mandate to the committee was clearly based on the premise that mental retardation as a problem depends upon input, involvement and action by all of the constituent agencies and could not be construed as the concern of any single agency.

3. Dr. Martha Eliot, then chief of the Children’s Bureau, in her report to Congress listed mentally retarded children as being in particular need of attention by this nation. While the Children’s Bureau had since its inception been interested in and concerned about the welfare of the mentally retarded, as evidenced by its pioneer studies of this group in 1913 and 1917, Dr. Eliot’s prescriptive response on the part of the Bureau in 1955 called for early detection, comprehensive evaluation, interpretation of findings to parents, and the use of such findings as a basis for ongoing help and care. This action recommendation not only highlighted a multidisciplinary approach but also visualized these children as requiring an intensification of the provisions made for all children for preventive health services, child health supervision and the building of good parent-child relationships.

4. To reinforce Dr. Eliot’s recommendations and to begin to implement the NARC proposals and Department of H.E.W.’s response, four special demonstration clinical programs were funded with MCH and CC funds. In these projects a real effort was made to explore and demonstrate how clinical services could be delivered by a multidisciplinary team within the program framework of the state MCH and CC services.

The congressional response to these major events over the next five years was an extremely positive and supportive one. Congressional intent was definitely in the direction of evolving a total balanced program which was multidisciplinary and multiagency in nature and which provided for access of the retarded child and his family not only to generic services which all families have a need to utilize at some point or other, but which provided authorization and appropriations for the constituent agencies of the Department of Health, Education and Welfare to initiate the specialized kinds of services required by this group. The appropriation and earmarking of MCH funds for the retarded, the authorization and appropriation of funds for the expansion of teaching and research in the education of retarded children, the funding of research in areas appropriate to the National Institute of Mental Health, the National Institute of Neurological Diseases and Blindness, etc., represent this congressional intent.

These beginnings served as the basis from which the President’s Panel on Mental Retardation developed its effort. As you may recall, in a spe-
cial message to Congress on October 11, 1961, President Kennedy established a panel of experts from various fields to make a broad study of the scope and dimensions of various factors relevant to mental retardation. The panel was charged with appraising the adequacy of existing programs and the possibilities for greater utilization of current knowledge. They were to explore the possibilities and pathways to prevent and cure mental retardation, as well as ascertain gaps in programs and any failure in coordination of activities.

The report of this group, presented to the President in October of 1962, calling for national action to combat mental retardation, is truly a remarkable document. While it calls for bold thinking and planning and for attacks on some of the root problems of our society as one way of preventing mental retardation, it also details, via some 94 specific recommendations, ways in which we can close gaps in service and improve the efficiency of programs as well as provide the resources to mount the program. The recommendations make repeated references to the necessity for training the multidisciplinary group of individuals required to deliver appropriate needed services in ways of working together—ways in which this group could be trained to meld their individual findings into a composite prescriptive program for the retardate and his family—a true team approach. The report suggested joint and interdisciplinary training as the approach which seemed most logical for achieving a mutual understanding, appreciation and coordination of disciplinary input into a total program.

These recommendations for multidisciplinary training were considered and proposed for inclusion in the legislative package which the administration prepared as the 1963 mental retardation amendments. Since the specifics of just how such interdisciplinary training was to be achieved were somewhat unclear, the Kennedy Foundation prior to the introduction of any legislation called together a number of university representatives for comment. While these representatives basically agreed that such interdisciplinary training could be mounted, they indicated that to do it well, consideration should be given to the provision of special facilities within which such training could be offered. Space was at a premium in most universities. Services were scattered and generally not proximate or related to teaching programs. If services could be assembled into a comprehensive demonstration model and if this could be incorporated into a physical facility which also housed the faculty and was designed to facilitate the learning process by students, the program could easily be launched.

Based on this input from universities, the initial phase in the development of multidisciplinary training programs evolved as Title I, part b of P.L. 88-164. This authorized federal support in the construction of facilities which
ultimately would house these training efforts. While the intent initially was to add some provisions for staffing the facilities to be constructed, this was dropped when the staffing provisions for the community mental health construction authority which constituted Title II of P.L. 88-164 ran into some difficulty.

The training programs in the to-be-constructed facilities from this point on were completely dependent on multiple sources of support which had to be melded and coordinated in much the same way as the proposed multidisciplinary training program ultimately would have to be evolved. It is interesting to note that the leadership in the planning process for the construction and eventual program which was to follow was assumed, in most universities expressing an interest, by an existing mental retardation clinical unit. These were basically the same clinical units which had been stimulated and supported as demonstrations by the maternal and child health program. They had the experience and know-how in the delivery of model comprehensive multidisciplinary services and a beginning exposure to the problem of training the people to deliver this service. The roots of the 1955 effort emerged clearly in this beginning.

In 1965, based on a review of the projected programs to be carried out in the facilities approved for construction under P.L. 88-164, it became evident that a major gap existed in the planned and hoped-for input of the staffing and operational support of the UAF centers from the constituent agencies of H.E.W. This gap related to input and support for the health services component of the UAF from the Maternal and Child Health Service. MCHS at that point had no specific training authority and certainly did not have the resources to support this needed component even though they had pioneered its development since 1955. As a result, a new section 516 was added to Title V of the Social Security Act. This section was subsequently changed to section 511, Title V of the Social Security Act and constitutes the bulk of the current MCHS investment in interdisciplinary training.

These health components constitute only one aspect of a total UAF program. The health component is essentially an interdisciplinary faculty and training program within a broader interdisciplinary setting. A minimum requirement is: full-time, visible, pediatric representation, pedodontic representation, social work, clinical psychology, special education, nutrition, physical therapy, occupational therapy, administration, speech and hearing.

*P.L. 89-97 authorizing "grants to public or other nonprofit institutions of higher learning for training professional personnel for health and related care of crippled children, particularly mentally retarded children with multiple handicaps."
public health nursing, and laboratory competency in the area of biochemistry and genetics. It is assumed that the mere existence of this kind of faculty would expose the students to some interdisciplinary concepts.

Your dental students, for example, by this faculty exposure would not only get some of the concepts in genetics that were presented this morning but also should learn something from the clinical psychologist in terms of behavior modification and the management of these handicapped children. They surely ought to learn something from the administrator in terms of how to manage the service delivery program. They should learn something from the social worker in terms of family relationships and history taking. Their association with a pediatrician, neurologist and psychiatrist ought to have some impact. They ultimately should be able to see themselves as a part of a total program effort and should be able to make their own unique contribution in concert with these other professionals.

A second major impact of the University-Affiliated Center programs relates to the academic-degree-granting departments. Each of these core faculty members in the UAF centers has a dual appointment—in the center as well as in a degree-granting professional school or department. This has several purposes. First it provides some assurance from the degree-granting academic department that the particular individual is considered by that department to be capable of teaching concepts as they relate to that profession. It also requires that the individual with such an appointment get involved with the departmental curriculum committee and do some teaching in exchange for the academic appointment. These interrelationships have had a real impact on undergraduate students and on the development and the content of the curriculum of the academic department.

In many of these university-affiliated centers a great effort has been made to develop what is being termed a “core curriculum.” With the kind of student mix in these programs it has been possible in those Centers to identify aspects of courses that are required by a number of professional schools and departments whose students are participating. It has been possible to define some content areas such as genetics, growth and development, management, etc., which are of interest and are required by a number of students from various disciplines. It has been possible to present that kind of content for credit to a variety of students within the UAF in a single course.

Interdisciplinary training, as I have indicated, is a newly-emerging concept. While it is still somewhat difficult to define, most of the UAF centers are somehow achieving parts of it. Even students who have gone through these programs, while they are not able to say exactly how they got some of their newly-found concepts, do seem to be getting them. This is the newer
direction which all of us need to explore, not only in terms of broadening the capability of our training products, but also in terms of achieving more efficient and effective use of our training resources, and ultimately being able to deliver a more meaningful total service to the handicapped child.
PREVENTING DENTAL DISEASE IN THE HANDICAPPED CHILD:

A CHALLENGE TO THE PROFESSIONS

Arthur J. Nowak, D.M.D.

Despite progress in the control of acute episodic disease in children through improved environmental hygiene, social and educational circumstances, antibiotics and immunizing agents, there has been little progress in the prevention of chronic disease and handicapping conditions—both mental and physical.

Programs for children with chronic disease present some difficult problems, e.g.,

1. The exact diagnosis of the disease is frequently difficult to determine;
2. Many children have more than one handicap;
3. A disease may cause a major disability in one child and a minor disability in another;
4. The disease process may improve with resultant lessened physical disability, or worsen with resultant greater physical disability.(1)

Estimates of the number of children with handicapping conditions vary from study to study and from group to group, which tends to complicate the development and planning of improved systems for delivery of health care to handicapped children (Figure 1).

There are many developments, social, economic and medical, that will effect the numbers of handicapped children in our country, e.g.,

1. The early detection of birth defects in the fetus through aminocentesis
Handicapping Conditions Among Children Under Age 21

Additional 1980 estimates:
Congenital heart disease 2,940,000; Emotionally disturbed 6,850,000;
Diabetes 122,000; Asthma-Hay fever 555,000.

is now an accepted component of good maternity care;
2. Family planning and birth spacing, if practiced by those who currently
do not practice them, will yield substantial child health benefits;
3. Limitation of abdominal radiographs and avoidance of unnecessary
drugs during pregnancy;
4. Comprehensive maternity and infant care to low income mothers;
5. Identification of “high risk” mothers and infants with careful manage-
ment prenatally and postnatally;
6. Mass immunization programs followed by systematic use of vaccines
in routine health supervision;
7. Screening programs to detect PKU, galactosemia and other inborn er-
rors of metabolism to allow for early treatment;
8. Early identification of childhood health problems that create handi-
capping states such as difficulties in vision, speech and hearing and
developmental delay.

These few examples and many others will continue to affect the popula-
tion of handicapped children. (1)

Unfortunately, though, there is a serious obstacle to identification of
health problems in the preschool child on a total population basis. That is,
only a small section of the child population receives continuous health care and supervision from infancy to school age. Those that are can be evaluated and early treatment begun, if found to have a handicap.

Because in the past in our society children were not brought together regularly in groups until school age, it was almost impossible to conduct screening examinations on this population. But it should become more of a reality in the future.

There is a trend in education now to begin children in the “learning experience” as soon as possible. Because of this philosophy there has been a growth of “early developmental type” programs established in many communities, not only in the suburbs, but also in the inner city.

Here the opportunity will be available for early screening of the children with the proper referral and follow-up. Also, children of “high risk” mothers can be followed and these parents encouraged to enroll their children in these programs at the earliest possible date. Here the health professional personnel and the special education teachers and therapists can identify those children with delayed or absent developmental signs and consult with the parents as to the most effective treatment and follow-up.

Once identified though, these children with delayed developmental signs have a multiplicity of needs in addition to the specific medical need of the particular condition or entity.

To meet this problem, comprehensive care clinics for the handicapped child have been developed in conjunction with university or health centers. These are integrated multidisciplinary efforts to meet the varied and complex needs of children with handicaps, usually made up of physicians, dentists, educators, therapists, psychologists and other allied health personnel.

But in numbers these are few and much of the diagnosis and treatment of the handicapped child still is carried out in the local community by the private general practitioners. Over the years they have performed to the best of their ability, but with the rapid development of new knowledge concerning health care for handicapping states, specialized care is now becoming available in increasing numbers in many communities, supported by both the governmental and private agencies.

All too often, in the case of dentistry, there has been a very limited amount of preventive treatment and management of dental disease in the handicapped child, leading to conditions that required the extraction of teeth instead of the repair of teeth and the prevention of dental disease. This has been demonstrated in studies (2,3) which report that the handicapped child has a very high DMF index (concluding therefore that he is more susceptible to dental disease), when, in fact, if one analyzed the DMF index, one finds that the decay rate (D) is quite normal, whereas the miss-
ing rate \((M)\) is high and filled rate \((F)\) is very low. This all supports the position that very little restorative and preventive treatment is available while exodontia is the dentistry of choice.

Where, then, can we as a profession provide major changes in the delivery of care in the area of prevention and maintenance of dental health in the mouths of the handicapped children, so that they will be able to develop to the best of their ability?

I would like to propose that we develop dental health programs, preventively oriented, to begin prenatally with parental counseling, followed by early examination, and then institute those preventive procedures that have been shown to effectively maintain the health of the oral structures.

**Perinatal care**

Prospective parents, especially those of first babies, are eager to have an adequate understanding of infants and children and of their care. Couples can be prepared for their roles as parents during the prenatal period by providing them with prenatal counseling. They should be taught what to expect when they become parents; their questions can be answered, and their fears can be alleviated. Finally, they can be made more confident of their own abilities and more aware of the community resources available to them. Prenatal counseling can be accomplished by either individual discussions or group conferences in conjunction with professional, educational and community representatives. (4)

Hospitals, service organizations, church groups and private health clinics may sponsor discussions. Allied health personnel should be included and here is where dentistry must get involved and take an active role.

With our increased understanding of the etiology of dental disease\(^{(5,6)}\) and with convincing evidence from laboratory research and clinical practice on what we can do to prevent disease, it is up to the dental profession to make this information available to the public, especially to couples about to experience the birth of their child.

Along with the usual topics of discussion in prenatal classes, the following dentally related topics must be discussed:

1. Development and maintenance of dental health in the parents' mouths;
2. The use of fluorides—systemic and topical\(^{(7)}\);
3. Development and eruption of teeth and occlusion;
4. Bottle habits, especially at night\(^{(8)}\);
5. Development of eating habits—frequency and quality\(^{(9)}\);
6. Effect of digital sucking and abnormal tongue placement;
7. Methods of cleaning the mouth, teeth and gingiva\(^{(10)}\);
8. Time of initial professional examination and consultation (11);  
9. Maintenance schedule to follow through the first fifteen years of life;  
10. Cost of dental care, including insurance coverage, government plans  
    and community resources.

**Neonatal care (birth to one month)**

Every infant should be under the continual supervision of a qualified  
physician. In those cases of a high risk infant the specialty services of a  
pediatrician should be requested. Dentistry will play a limited role during  
the neonatal period. The only instance may be with the supervision or re- 
moval of a neonatal tooth or in the examination and consultation of a con- 
genital anomaly.

**Postnatal care**

Based on current practice and knowledge the following schedule of visits  
is recommended by the Academy of Pediatrics (4):  

1st year, 6-9 visits;  
2nd year, 2-4 visits;  
3rd year, 1 visit;  
Through adolescence, 1 visit per year.

Such a schedule may not be appropriate for all situations and carefully  
considered modifications can be made with regard to the circumstances in- 
volved.

Naturally the physician and his staff will be responsible for the medical,  
emotional and psychological examination of the child. In too many cases,  
though, the physician at his present level of training is not competent to  
effectively examine and consult with the parents as to the dental health of  
the child. (12, 20) Also, the suggested schedule of preventive child health  
care developed by the American Academy of Pediatrics is in error as to  
the time of the initial examination of the child's mouth by the dentist. They  
state that a dental referral should be made at 2½ years, and we know that  
epidemiological studies (13, 14) show that by this age, almost 50 percent of  
the children have evidence of dental disease in the form of caries and mar- 
ginal gingivitis. But they do recommend some very positive preventive prac- 
tices before this age and I would suspect that they feel the physician and  
his staff are responsible for their development.

I would rather suggest that the child, with his parents, be seen by a den- 
tist no later than one year after the first tooth erupts into the mouth and I  
would further propose that the dentist and his staff be responsible for all
those preventive measures that the Academy recommends before the first
visit and thereafter.

In the case where the infant is of the high risk type or already is identi-

cified as an exceptional child, or if the infant is a product of a couple whose
dental history has been very poor, earlier initial examination and preventive
care is necessary, I would say no later than six months after the first pri-
mary tooth erupts.

**Personnel and time requirements**

The tradition of private dental practice has been that the dentist be pres-
ent at each visit. However, if all the children in the country are to receive
preventive care, many visits may have to be conducted by other dental
health personnel who have been trained in the early recognition of dental
disease and in those procedures, both educational and clinical, that have
been shown to be effective in its elimination and control.

In all cases though, a dentist should be available for further consultation
and evaluation and to supervise the auxiliaries in their responsibilities.
Every effort should be made to minimize the number of appointments and
reduce the parents' traveling time.

Also, it is important that all learning experiences in the office be backed
up by appropriate audiovisual aids and take-home materials that can be
prepared by the dentist and his staff to offer a more personalized approach.

The initial examination, which is performed by the dentist and followed
by a consultation with the parents, should require about one-half hour of
time. Any preventive instructional sessions are then handled by the appro-
priate auxiliary and will require fifteen to thirty minutes of time. Naturally,
those infants and young children with complicated histories and compli-
cated dental conditions will require longer appointments and more frequent
appointments.

Referral to specialists, especially pedodontists, should be suggested by
the family dentist in certain high risk cases and when exceptional children
require extensive treatment and hospitalization that the family dentist may
not be inclined to become involved with.

**Report of a pilot program**

The United Cerebral Palsy Association of the Pittsburgh District provides
direct services to the developmentally disabled with special emphasis on
cerebral palsied individuals and their families who reside in Allegheny
County. These services include:
1. *Early identification and treatment program* for infants up to age three who are evidencing developmental lags;
2. *Developmental centers* for children ages two to six who are being prepared for placement in school programs;
3. *Day care centers* for children ages six to seventeen who are severely physically involved and profoundly mentally retarded.

Because of the type of services provided, the population of cerebral palsy children naturally ranged from infants to late teens. This provided us with a full range of conditions to investigate—the infant’s edentulous mouth, the preschooler with complete primary dentition, the school-age child with a mixed dentition and the teenage child with an almost complete adult dentition.

It was decided, after lengthy discussion, that the Association was in no position to provide extensive dental care to its patients, but that it could provide educational opportunities to parents, early examination of the children, daily plaque control in the centers, modification of eating habits at the centers and daily application of topical fluorides. Those children that did require professional treatment could be referred to clinics or private practitioners that accept handicapped children.

The dental consultant, along with the executive director, developed a proposal (Table 1) for the program that was presented to the Board of Directors for its approval and implementation. It was the feeling of the dental consultant that this preventive program had to be first accepted by the Board, the staff and then the parents, to be effective, very much like the method of acceptance as proposed by Barkley. (15)

Once approved, a series of presentations and workshops was conducted for the staff on the problems of dental disease and its control through plaque removal, modification of eating habits and multiple use of fluorides. They began immediately to practice these procedures in their own mouths, and after one month, another workshop was conducted to review the procedures and answer any questions. The staff was then asked for their acceptance of this program and their cooperation in its implementation. We received their unanimous approval to go ahead with the children.

A series of two “parent night” presentations was held at the downtown center to inform them of the program and to seek their support and cooperation.

To implement the programs at the centers, it was felt that the supervisor at each center would act as team leader and design the program with her staff in the manner that best suited their individual needs, always remembering to base it on the principles that had been laid out for the total pro-
Development of a Preventive Dentistry Program
for the Young Cerebral Palsy Child

Purpose: To prevent dental disease from developing in the mouths of the young cerebral palsy population by teaching the professional staff and parents proven plaque control techniques

Facility: United Cerebral Palsy Developmental Facility

Population: Cerebral palsy children
a. 100 children
b. Ages 2.6-6.6 years old

Method:
1. Vote of approval and acceptance
   a. Executive Director
   b. Board of Directors
   c. Dental Consultant
2. Educating the professional staff
   a. Workshops
   b. Demonstration
   c. Participation
3. Informing the parents
   a. Parents night
   b. Demonstration
   c. Acceptance
4. Designing the program
   a. Professional staff (teachers, therapists, nurses)
   b. Parents
   c. Executive Director

Program:
1. Elimination of plaque
   a. Toothbrushes—manual and electric
   b. Flossing—holders
   c. Staining material—liquid
   d. Mouth props
   e. Grading system
2. Diet counseling
   a. School snacks and meals—dietician
   b. Home meals—parents
3. Fluoride therapy
   a. Supplements—diet
   b. Home application
   c. Professionally applied
4. Dental examination
   a. Radiographs
   b. Restorative treatment
   c. Sealants
gram. It was also suggested that they start with one technique first, for example, toothbrushing, and not proceed to another until they could all master that procedure.

While the supervisors were designing their programs, all the children received a dental examination with a light, mirror and explorer. Saliva samples were drawn for the Modified Snyder's Test(16) and a plaque score, based on the Simplified Green-Vermillion technique(17) was given. Parents were encouraged to accompany their children for the examination that was conducted at each center so that the dentist could consult with the parents as to the individual needs of their child and answer any questions that they might have. Also, it was then possible to receive additional information as to the home care procedures, eating habits and dental attitudes of the parents. All this material was gathered and is being used as our baseline data for future comparison.

Results of the examination were typically what one would expect. The young infants had no carious defects but evidence of marginal gingivitis. The preschool children evidenced incipient carious lesions and marginal gingivitis, and as the patient progressed in age, so did the defects of dental disease.

Information gained from the parents as to home care procedures showed that the majority of the children never have their teeth brushed, are not using fluorides, have never been to the dentist and eat a predominantly carbohydrate diet of the sticky and sucrose variety and expose themselves to this type of food at least three to six times a day.(21)

For the first three months of this program the children's teeth were brushed daily by the staff after meals, with weekly plaque checks using the liquid staining material. At the same time, every attempt was made to eliminate sugary snacks and meals heavy in carbohydrates. Conferences were held with the parents to encourage them to do the same at home.

After three months the staff felt comfortable with the brushing techniques and other procedures were now introduced. The use of floss for interdental cleaning with or without holders was found to be most difficult but, nevertheless, in most of the children a daily attempt was made.

We knew most of the children were not on fluorides, and so, recognizing the beneficial effects of daily applied fluoride to the teeth as reported by Ericson and Foreman(18) and Shannon(7), a letter was sent to all parents requesting permission to brush on a fluoride gel (1.23 percent NaF in a low pH base) after the tooth cleaning was performed. This procedure has been found to be quite routine and has encountered very little objection from the staff or children.
**Preliminary observations**

Although the program has been in effect for only seven months, one can sense and witness the overall improved dental attitude of the executive director, the staff, the children and the parents.

Follow-up dental examinations revealed improved oral conditions with much improved plaque scores, lower Snyder Test scores and evidence of dental restorative treatment.

There has been a decrease in sucrose-type snacks and an improvement of the quality of the lunches. The children are familiar with the toothbrush and floss and even are attempting to clean their own teeth.

The staff has devised methods of positioning the children in a supine position, for better control and access. They have found the manual toothbrush to be satisfactory and do not care to get involved with electric brushes.

Parents are enthusiastic and are now seeking out further professional care so that the other preventive measures may be made available for their children. A number of the more severely handicapped children with numerous dental problems have undergone full mouth rehabilitation at hospitals and follow-up preventive care can easily be maintained at the centers.

All new children accepted in any of the programs of UCP are examined by the dentist and immediately placed in the Preventive Program. His or her parents are taught the home care procedures and given information as to eating modifications and reduced exposures.

**Long term results**

The final results of an early identification and examination program, followed with daily preventive procedures, obviously will require observation for a number of years before one can determine its overall effectiveness. However, one must believe that, with the improved oral ecology along with multiple applications of fluoride and reduction of sucrose intake, the oral health of the handicapped child will compare with the results one can get in a normal population. Because the program is based on sound scientific procedures that have been proven effective and initiation of the program is at a very early age before the disease process can take hold, we should observe significant reduction in dental decay, gingival inflammation and loss of teeth.

But it can only be effective as long as we have the cooperation of the staff, the parents and the health professional team of physicians, dentists, nurses and dental auxiliaries.
Continuing education programs for these groups, along with periodic workshops for upgrading information, discussing mutual problems and understanding of mutual problems, can only improve communications and motivation, as well as the success of the program.

Summary

There is sufficient evidence before us that with improved medical technology, birth control methods, family planning and early identification of birth defects, the number of handicapped children will be affected. There is also a move on in the country for education of all children, including the handicapped, and beginning at an earlier age than previously customary. There is also sufficient evidence that dental disease can be prevented and healthy mouths maintained in all patients. Therefore, it is suggested that the dental profession:

1. Become involved in prenatal courses and design presentations to create an awareness in the new parents as to the need for early preventive dental practices;
2. Inform the Academy of Pediatrics that the initial examination of the mouth of the child should be performed by 18 months of age and that parents should be taught the home care procedures of plaque removal, fluoride therapy and diet modification by the dental team;
3. Become involved in all educational programs for the handicapped and develop and institute a preventive program based on proven techniques;
4. Continue to research the various methods of prevention now being used in the country and investigate their effect on the handicapped child.
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A COMPREHENSIVE CLINICAL DENTAL PROGRAM FOR HEMOPHILICS

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One of the most neglected aspects of care for the hemophiliac is proper dental treatment. Many dentists are reluctant to treat hemophilic patients and parents of hemophiliacs often unwisely postpone needed dental care. It is unfortunate that dentistry is often relegated to secondary importance, since this neglect may pose future problems which may give rise to life-threatening crises. (10)

Dental caries is still one of the major diseases in the world. It affects 93 percent of the population of the United States. (9) The child with hemophilia is unfortunately not free of this disease. In fact, the incidence of dental disease is equal to or surpasses that in the normal child. Current knowledge of the etiology and pathogenesis of dental caries has made possible more effective methods of caries prevention and control. Properly applied, this knowledge could eliminate one of the economic burdens facing the hemophiliac or the parents of hemophilic children.

A thorough review of the literature reveals that most papers deal with oral surgery and case reports. Few articles are devoted to the comprehensive dental care of hemophiliacs. The purpose of this report will be to outline the principles and techniques for a comprehensive clinical dental program for hemophiliacs.

Dental care for the hemophilic child should begin before the age of three years. A detailed medical and dental history must be obtained, along with full mouth roentgenogram, study models and photographs and an oral ex-
amination should precede the formulation of a diagnosis and treatment plan. Psychological evaluation of the patients should be secured whenever it is felt that this information would be an important factor in the overall plan for dental treatment. A case presentation to the hemophilic patient and his parents, with special emphasis on preventive measures, is essential. In a dental health care program for hemophiliacs, strong emphasis must be placed on a multidiscipline team approach.

Since most dentists do not have the special medical expertise necessary for dealing with hemophilia, the cooperation and close collaboration of a physician with appropriate experience in managing hemophiliacs are essential requirements before undertaking dental treatment. The dentist providing the care must understand the nature, manifestations and overall treatment of the disease and associated psychosocial problems. The multidisciplined approach of dentists, physicians and paramedical personnel involves consultation for every patient before and during the course of dental treatment. The utilization of modern medical knowledge and treatment techniques is one of the two factors which make dental treatment of the hemophilic child feasible; the other is the utilization of advances and refinements of modern dental science.

**Classification**

The most common type of hemophilia is hemophilia A or classical hemophilia, in which the protein factor missing is an antihemophilic factor (AHF, or factor VIII). Approximately 75 percent of all hemophiliacs are type A. The second most common variety of hemophilia is hemophilia B, or Christmas disease. The protein factor missing in hemophilia B is Plasma Thromboplastic Component (PTC, or factor IX). The third classification, hemophilia C, is an extremely rare form of hemophilia. (2) Each type of hemophilia is further classified as mild, moderate, moderately severe or severe.

In a given family, the type and severity are the same. For example, in a family with classical hemophilia, all affected males will have approximately the same factor VIII level. In one family, all affected males may have severe hemophilia (1 percent or less factor VIII level), while in another family, affected males may have moderately severe hemophilia (1 percent to 5 percent factor VIII level). In a third family, affected males may have moderate hemophilia (5 percent to 25 percent factor VIII level). When there is 25 percent to 50 percent of the missing factor present the hemophilia is considered to be mild.
Mutations

A hemophiliac may be born into a family in which there is no history of hemophilia. If the mutation occurs in an ovum which becomes a male, hemophilia will be manifested in the male newborn. If the mutation takes place in an ovum which becomes a female, she will be a carrier. This carrier state, however, may not be evident until she bears a hemophiliac. The mutant gene may pass through several generations of females before manifesting itself in a male.

Andonian, Dietrich and Whiteman (1) found that approximately two-thirds of their Hemophilia Project's 500 patients knew of other hemophiliacs in their families. In the one-third without such positive histories, we can assume that a mutation or change had occurred in the gene of the mother.

The following is a brief outline of screening tests for determining the presence of a bleeding problem.

1. Capillary bleeding time: Test for integrity of vascular and capsular system; normal time is one to three minutes.
2. Platelet count: Normally 2,000 to 4,000 per cubic millimeter; evaluates platelets for integrity.
3. One-step prothrombin time: Normal is 11 to 15 seconds:
   A. Picks up deficiency in step II (prothrombin to thrombin) of coagulation;
   B. Detects disorders in this stage but does not give a definite diagnosis;
   C. Inversely proportional to prothrombin content of plasma.
4. Partial thromboplastin time (PTT):
   A. Normal is 30 to 45 seconds;
   B. Most useful screening test for blood factor deficiency in stage I (thromboplastin formation).

The above four tests (4) and an adequate history should disclose disorders. (12)

Clotting factor levels in carriers of hemophilia

The average factor VIII level of carriers of hemophilia A is about 50 percent, with a range from a few percent to well over 100 percent. That is, about 50 percent of carriers have factor VIII levels below normal and the remaining 50 percent have levels overlapping the normal range.

Carriers of mild hemophilia may sometimes have low levels of clotting factor. Estrogen and progesterone, in combination and in sufficient dosages, will elevate factor VIII and factor IX levels in normal females or carriers. Therefore, females should not be tested for an estimate of their carrier status while they are pregnant or taking contraceptive hormones. Carriers with low factor VIII or factor IX levels, who ordinarily bleed excessively at trauma or operation, may bleed less when placed on contraceptive hormones.
and may have less bleeding at delivery than one would expect from their nonpregnant clotting factor levels or menstrual histories. Carriers with low clotting factor levels need the same care as males with mild hemophilia with similar clotting factor levels.

**Inhibitors**

Inhibitors are found in about five percent of patients with severe hemophilia A and very rarely in patients with severe hemophilia B. These patients have developed an antibody (inhibitors) to factor VIII or factor IX. The etiology of inhibitors is undetermined. Patients who develop a high titer of inhibitors usually do so in early childhood, often after a few transfusions of plasma. Lower titer inhibitors usually occur for the first time in late childhood or adulthood.

When a patient with an inhibitor is transfused with plasma concentrate, the clotting factor molecules will combine with antibody molecules to become inactivated. If enough plasma concentrate is given and the antibody titer is not too high, all the antibody may be used and any factor given in excess of that point will remain in the plasma and be able to participate in coagulation. If a patient who already has an inhibitor receives a dose of plasma concentrate, the clotting factor in the plasma may stimulate even more inhibitor production. This is especially true in patients who tend to have high titer inhibitors; the effect may be minimal in patients with low titer inhibitors.

Plasma concentrate must be withheld from patients with inhibitors, especially those with high titers, to avoid stimulation to higher titers. Even very high inhibitor titers will gradually decline if the patient is not exposed to the antigen. If bleeding occurs, local hemostatic measures such as pressure and cold are used. Prednisone is used for a period of a few days. If the hemorrhage is very serious or life-threatening, a high dose of plasma concentrate sufficient to overcome the inhibitor may be given. Plasmapheresis may be employed first in order to eliminate some inhibitor-containing plasma.

**Hepatitis**

All blood products useful in treating hemophilia can transmit hepatitis. Severe hemophiliacs who receive continuous transfusions of blood products are usually exposed to hepatitis. Some develop clinical hepatitis; most do not. It is the authors' opinion that the severe hemophiliac must risk the chance of hepatitis in order that the required hemostasis is achieved for optimal dental care.

Every consideration, however, should be given to avoiding transfusions to
patients with moderate or mild hemophilia, who seldom have a serious bleeding problem. Minor bleeding may respond to local measures. Superficial operative procedures in nonvital locations may be performed without plasma products if the surgeon is attentive to bleeding points. Women can be placed on contraceptive hormones to elevate clotting factor levels.

Allergic reactions

Patients have far fewer and milder reactions to cryoprecipitate or plasma concentrate because these contain much less extraneous protein. The manifestations of allergic reactions are hives and chills. To counteract these reactions, large dosages of antihistamine are often given.

Dental anesthesia

In the dental treatment of hemophilic patients the use of any type of anesthesia should never be considered as a routine procedure. When anesthesia is indicated, each hemophiliac must be evaluated on the basis of previous medical and dental history before determining the most suitable anesthesia.

For regional block anesthesia, a factor VIII or IX level of 30 to 50 percent is obtained pre-operatively, and a level of 20 to 30 percent maintained for 24 hours, with a daily postoperative examination for seven days. If during aspiration a blood vessel is injured and blood is aspirated, the patient should be maintained at a 20 to 30 percent factor VIII or IX level for 36 to 48 hours. For infiltration anesthesia one infusion to raise the factor level to 20 to 30 percent is given pre-operatively. The severe patient should be monitored for five days. The necessity of plasma products therapy is debatable in the mild or moderately severe hemophiliac.

For patients with severe hemophilia undergoing general anesthesia, plasma products should be given to obtain a pre-operative factor VIII or IX level of 50 percent or above; a level of 20 to 30 percent is maintained for 24 hours postoperatively. The intubation should be oral to protect the nasal conchas and adenoid tissues from trauma and to prevent bleeding into the nasal area. It is often difficult to assess the extent of damage when bleeding occurs into the nasal area.

General anesthesia with intubation may be preferred in cases necessitating multiple extensive restorations (4), since the patient would be exposed to fewer bleeding risks and medical complications. The cost of repeated replacement therapy for block anesthesia versus the one time for general anesthesia is also a consideration.

When a short duration and light plane general anesthesia is used in combination with a local anesthetic for oral surgical procedures, oxygen is ad-
ministered for two minutes, during which time an intravenous infusion is established. Methohexitol sodium, one percent solution, is administered in a dosage sufficient to induce sleep. When sleep is induced, two percent lidocaine hydrochloride with epinepherine (1:1,000,000) is forcibly injected into the pericemental space around the teeth using a 29 gauge needle. After surgery, the patient should be placed on a soft food diet until he can return to have the occlusion and marginal ridges checked and all high spots removed. Nitrous oxide analgesia is a particularly useful technique for achieving a comfortable dental experience.

Hypnosis has been used as an adjunct to anesthesia or as a substitute when block anesthesia is contraindicated in patients subject to hemorrhages not readily brought under control. (7,8)

Psychological behavior

It is not uncommon for the hemophilic patient to be erratic in keeping his appointments and to behave suboptimally in his conditioning and receptiveness to the dental treatment. A possible explanation for this behavior lies in numerous problems which confront these patients. Mattsson and Gross (10) reported that the dentists and physicians who are treating hemophilic patients must be familiar with the associated emotional problems accompanying hemophilia. They should also be especially alerted to the frequent guilt feelings and over-protective nature demonstrated by parents of hemophilic patients. Complex emotions demonstrated by the multihandicapped hemophiliac sometimes make it most difficult for him to adjust and accept dental treatment.

Some studies have evaluated the relationship between psychological and educational problems in the hemophiliac. Taylor (15) has described how the repeated interruptions in school attendance due to bleeding episodes, etc., result in many patients feeling hopelessly lost academically by the time they reach adolescence. The hemophiliac may be placed in a school for handicapped children, where less is expected of him academically. His ultimate educational achievement is apt to be substandard, despite his normal or superior intelligence. He may continue to lead a sheltered life, with a closely-hovering mother. His father may discourage him from participating in sports and other masculine pursuits and he may become alienated. The entire family may suffer from the stresses and strains of this chronic, painful, emotional and extremely expensive illness, which requires so much of the mother's time and energy.

The stress of having hemophilia, added to the normal stresses of adolescence, often leads to the eruption of over-emotional problems in teenagers. Boys who have become crippled are especially apt to become withdrawn
and depressed. Those whose mobility is normal or near-normal may revolt, showing wild and careless behavior. Some patients may become habituated to analgesic drugs. The dentist may find it necessary to give special consideration to arranging treatment time for hemophiliacs in order to minimize school absences and loss of time from work. He may also find it necessary to alter the way in which he would normally communicate with the patient.

The patient who reaches adulthood with severe deformities, minimal education, an immature personality or a combination of these leads a dependent and unrewarding life. He may be educationally unqualified for any job other than physical labor, which is unsuitable. The implications of these problems to total dental care for the most part have been overlooked. Other hemophiliacs, who have had appropriate medical treatment, adequate vocational training and family environments that have been emotionally supportive may have minimal disabilities on reaching adulthood and find a satisfying career. Patients with moderate or mild hemophilia tend to lead more normal lives.

Oral surgery

Oral surgery should not be performed until the patient has been examined and cleared by a hematologist or physician knowledgeable about hemophilia. A decision should be made regarding the institution of replacement therapy and chemotherapy and the duration of this therapy. The patient should also be tested for inhibitors.

Arrangements to administer plasma products and other medications at either the office, clinic or hospital are required.

In some patients, notably those with Von Willebrand's disease, the bleeding tendency also appears to go through phases in which the hemophiliac responds differently. This variability in the patient's bleeding tendency is of the utmost importance in scheduling dental treatment. Extraction of teeth is delayed whenever possible until the patient is in a quiescent bleeding period. The usual dosage for the severe hemophiliacs is to raise the deficient factor level to 20 to 30 percent one hour before the extraction. A repeat dose of plasma product is given only if rebleeding occurs. For very extensive oral surgical procedures, plasma products may be given twice a day for seven to fourteen days, to maintain a minimum factor VIII or IX level of 20 to 30 percent during a prolonged healing period. Epsilon-amino-caproic acid is begun on the day of extraction and continued for days. Mild to moderate hemophiliacs usually have uncomplicated extraction of permanent teeth if given plasma products preoperatively to a 20 to 30 percent level.

Local anesthesia by pericemental injection along all four quadrants of the
tooth is used for extractions. Block anesthesia is inadvisable and not necessary. The tooth is extracted with as little trauma as possible. Following extraction, Surgicel® and thrombin powder are placed in the socket and gauze pressure packs applied. Splints are not used. Patients are instructed to take nothing except the EACA suspension by mouth for 24 hours. Cool, clear liquids (no dairy products) are permitted during the next 48 hours. For the following seven days, pureed food or food not requiring mastication is permitted. The patient contacts the surgeon’s office once a day during the ten days after the procedure.

Exfoliation of primary teeth

Exfoliation of primary teeth extends over a lengthy period and is sometimes accompanied by periodic or prolonged oozing of blood. (6) When oozing of blood is persistent, it is advisable to administer appropriate plasma products, to correct the patient’s hemostatic defect, and remove the tooth or root tip. If a local anesthetic is required for extraction, pericemental injection is used, and all the pulpal tissue beneath the primary tooth is curetted in order to promote good healing. Hospitalization is not necessary, but the administration of plasma products and/or epsilon-amino-caproic acid may be appropriate. The need for these agents, and the dosage and duration of administration, is determined by considering the severity of hemophilia, the length and number of roots to be extracted, the number of teeth to be removed at one session, the extent of trauma anticipated, and the presence or absence of infection. A mild hemophiliac may require no plasma products or other medication for extraction of a partially-exfoliated tooth. A severe hemophiliac may receive plasma products to raise the plasma level of his deficient factor to 20 percent or more for an uncomplicated extraction. For difficult, complicated or multiple extractions, a plasma clotting factor level of 30 percent or more should be achieved before the procedure, and epsilon-amino-caproic acid therapy instituted. A single dose of plasma product immediately prior to the extraction is usually sufficient, but if rebleeding occurs hours or days after the extraction, another similar dose should be given.

Restorative procedures

Restorative procedures should not be delayed because the patient is hemophilic. Whenever possible, restorative procedures are performed without regional block anesthesia or local anesthesia. It is the authors’ opinion that primary teeth especially can be restored without anesthesia, for many restorations are done in this manner on normal patients.

Bleeding from the tongue is very difficult to control in a normal patient;
in a hemophiliac it can be a serious problem. Andonian, Dietrich and Whiteman (1) reported that hemorrhage in the lax tissue beneath the tongue may dissect into the anterior nuchal muscles or posteriorly to the pharynx. This is potentially dangerous, as airway embarrassment may occur. They stressed that one of the early signs of impending neck bleeding is a hematoma under the tongue. If this should occur, the patient should be immediately hospitalized. Control of bleeding from the lips and other soft oral tissues is difficult. Injury to such structures during operative procedures can best be eliminated by the use of the rubber dam which protects the tongue and oral tissues, provides the patient with a sense of security, and minimizes chances of accidental injury.

The rubber dam should always be used to restore in the mandibular arch to prevent possible trauma to the tongue. Whenever dental procedures which may produce bleeding are anticipated such as during the removal of gross caries extending below the gingiva or when a stainless steel crown is placed for a severe hemophiliac, replacement therapy should be instituted preoperatively, and occasionally maintained postoperatively.

The proliferation of gingival tissue may extend into the proximal areas of grossly carious teeth. A zinc oxide eugenol dressing may be placed in an attempt to facilitate resorption of this hypertrophied tissue to minimize hemorrhage during restorative procedures.

As in restorative procedures on normal patients, air rotors, rubber dams and clamps, stainless steel matrices and wooden wedges are necessary for the placement of quality amalgam restorations. When using air rotors, a new burr and wash-filed technique should be employed to minimize discomfort to the patient. The slight amount of gingival bleeding which may be encountered with the placement of rubber dam clamps, stainless steel crown, matrices and wooden wedges is usually not a problem. A rubber dam clamp such as the S.S.W. #26 is more suitable than the Ivory-14 type because it has no sharp points and reduces the possibility of puncture wounds during placement.

Stainless steel crown preparations are modified so that the finish line is above the gingival margin. The proximal reduction is made with a fine fissure burr, working from the tooth outwardly below the contact. Careful attention to minimal tooth reduction and careful crown adaptation significantly reduces the possibility of a bleeding crisis. In the placement of the stainless steel crown, the blanching of the gingival tissue is the best guide to removal of the excess, while fitting the crown frequently. One must remove the excess from the crown to prevent a future bleeding problem. If hemorrhage occurs unexpectedly, plasma replacement therapy should be initiated to raise the clotting factor level to 20 to 30 percent. Usually one
infusion will be sufficient to provide hemostasis and a good healing site. Prophylactic odontotomy practiced routinely obviates the need for restoring deep carious lesions with its accompanying discomfort at some later time.

Periodontal disease and prophylaxis

Because of poor oral hygiene and neglect of the oral cavity with the subsequent accumulation in bacterial plaque of materia alba and calculus, some patients should be given prophylactic doses of missing factor concentrate before, as well as following, periodontal treatment. Oral or intravenous premedication should be used when indicated, often in conjunction with replacement therapy. The commonly accepted procedures of scaling, curettage and polishing can be performed without blood products. However, care must be taken at all times to avoid excessive trauma. The sessions should be short and frequent with minimal bleeding and should allow for gradual healing. If undue bleeding or trauma to the gingiva is anticipated, such as during removal of large amounts of subgingival calculus with resultant laceration of the gingival tissue, or with chronically inflamed gingiva that are edematous and hyperemic, one preoperative infusion of blood products should be administered to raise clotting level to 20 percent. Periodontal surgery should be performed conservatively, keeping in mind the need of infusion, as in any major medical surgical problem. The patient's clotting factor level is elevated to 30 percent and is maintained at this level for at least three days or until signs of trauma begin to disappear and good healing has occurred. At the last appointment, patients receive fluoride treatment and all restorations.

The importance of good home care instruction cannot be overemphasized. This should include tooth brushing instruction, implementation of a daily flossing routine and stressing the importance of routine maintenance of all restorations. Many practitioners feel the incorporation of daily fluoride mouth rinses is a useful procedure in preventing unnecessary dental breakdown.

The use of water picks and electric tooth brushes as devices to help improve the home care techniques has produced excellent results, especially in those hemophiliacs who demonstrate dexterity problems or are undergoing orthodontic treatment.

Pulpal therapy

Both direct and indirect capping techniques have been used on primary and permanent teeth in hemophiliac patients. The indirect capping technique is ideally suited for hemophiliacs, since removal of most of the
carious dentin can be accomplished prior to exposure of the pulp. Since this procedure is well tolerated by the patient with little or no pain, a local anesthetic should be avoided. Should the indirect capping technique fail, the patient may later have endodontic therapy with plasma replacement.

Calcium hydroxide paste is placed over the remaining carious lesion, followed by a zinc oxide eugenol dressing, and the tooth restored. This procedure allows the tooth to deposit secondary dentin between the pulp and the carious lesions, which allows the pulp to regain a healthy state and remain vital. This procedure has been well tolerated by our patients, with little or no pain involved. Local anesthetic is unnecessary. In the event that the indirect capping treatment fails, the tooth is scheduled for endodontic therapy. Many clinicians recommend that a tooth with an indirect pulp cap should be reopened in about three months to remove any remaining caries and to observe if healing has occurred.

The use of formocresol pulpotomies in primary teeth of hemophiliacs is encouraged. Both the one-step and two-step formocresol pulpotomy may be performed with excellent results. This procedure has been employed successfully on teeth which are totally necrotic and involve a fistulous tract. An anesthetic solution may be injected directly into the pulp following exposure of vital pulp whenever necessary, whether or not the patient has received prior plasma therapy. This is ordinarily sufficient to anesthetize the tooth sufficiently to complete the pulpotomy. A cotton pellet moistened with formocresol applied to the bleeding tissue within the canal is sufficient to provide hemostasis without prior plasma therapy, making replacement therapy unnecessary.

Droter (5) reported using the two-appointment formocresol technique in 212 nonvital primary teeth without fistulas; only ten failures resulted. Due to the lack of adequate studies to evaluate this technique for permanent teeth, it is presently advocated only for primary teeth. However, some clinicians have used this technique in the treatment of badly involved permanent teeth as an alternative to extraction. (16) Conventional endodontic therapy is indicated on all permanent teeth if the root apices are closed and it is economically feasible. (14) Following the second step of the pulpotomy procedure, the tooth should be prepared for a stainless steel crown to insure against failure due to fracture or leakage.

Direct pulp capping and calcium hydroxide pulpotomies of primary teeth (using either calcium hydroxide or zinc oxide and eugenol), although still employed, are not as widely used as in the past. Many clinicians (11) (personal communications, pedodontic staffs, USC and UCLA dental schools) feel that direct pulp capping of primary teeth should be limited to minimal mechanical exposures and that the formocresol pulpotomy is
preferable due to its higher percentage of clinical success.

In direct pulp capping or pulpotomies on permanent teeth, anesthesia can be given by two methods. When replacement therapy is given pre-operatively, a local or general anesthesia may be used. (4) An alternative method is to inject the anesthetic solution directly into the exposed pulp, creating effective anesthesia and controlling bleeding through the vaso-constrictor present in the anesthetic solution. (4)

**Orthodontia**

Due to the general lack of knowledge concerning orthodontics for the hemophiliac and the frequent necessity of bicuspid extraction, many hemophiliacs have been denied the esthetic, functional and psychological benefits derived from orthodontic treatment. Malposition of teeth may lead to severe periodontal disease as well as caries.

Both orthodontics and interceptive orthodontics can and should be done for hemophiliacs. A chief concern is that the operator should be careful to prevent wires and brackets from irritating the soft tissue, since the resulting bleeding may be difficult to control. The prolonged use of bands may cause chronic irritation of the gingiva with resultant hemorrhage. To alleviate this problem the operator might consider reduction of the width of the bands so as not to traumatize the gingiva unduly. Removable orthodontic appliances when appropriate are ideally suited for the hemophiliac, since there is minimal irritation to the gingiva. In addition, periodic x-ray examinations may be performed without interfering with treatment.

**Summary**

It has been shown that with consultation with the patient's physician and using standard operative procedures with precautionary care, hemophiliacs can be treated, and dental health can be achieved in a hemophiliac. A comprehensive program will minimize the necessity for extractions, and the subsequent sequelae of tooth migration, periodontal disease, extraction of other teeth and prosthetic appliances. Conjoined with the medical approach, dental surgery can often avoid or minimize hospitalization.

Two fundamental statements summarize the treatment plan for hemophiliacs: for hemophiliacs, as for all, there is no substitute for good dental procedures, and dental treatment is not elective but mandatory; current knowledge of procedures and techniques for the prevention and treatment of the hemophiliac's dental ills no longer justifies the dentist's hesitation in treating them.

The hemophiliac has the usual dental problems found in the general population. His dental morphology and anatomy are clinically the same as a
“normal” patient’s, and so may be his dental history, if it is one of neglect ending in acute dental decay or disease because he fears bleeding, pain, possible hospitalization and loss of time from work or school.

Dental treatment for the hemophiliac is most effective as an integral part of a total rehabilitation program because the dental staff then has access to the physician, to the hospital, and to hematological and psychosocial services.

It is felt that in order to establish safe outpatient care for the hemophiliac, the technique and conservative methods of dental treatment presented in this study need to be joined with medical and psychological approaches and with patient education and cooperation. Dental procedures, especially extraction, should no longer be considered a life-threatening situation. The development of programs to treat hemophiliacs on either an outpatient, home care or private basis is a step in the right direction. The financial burdens, time lost from work or school because of hospitalization, and the emotional and psychological problems with dental treatment warrant the development of such programs.

Observations of hemophilic patients by the authors showed that a hemophilic patient postpones dental treatment partly because of his own ignorance of the need for dental care, partly because he fears ignorance on the part of dentists in the matter of treating hemophiliacs, and partly from discouragement about the prospect of losing more of his work or school time, continued disabilities and hospitalization.
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11. Personal Communication, Pedodontic Staff, USC and UCLA dental schools.


There are three separate but interrelated elements in the title of this presentation that are germane to the purpose of this conference. What follows is an examination of these elements based on a continuing experience in interdisciplinary enterprise which began in 1949 and includes observation of similar units in this country and abroad.

The first element has to do with problem solving in dealing with patients presenting craniofacial anomalies. Such anomalies include malformation of structure and abnormalities of function, growth, behavior and biochemistry. The problems encountered in this region, whether at the clinical or laboratory level, are so complex as to be refractory to solution by conventional unidisciplinary approaches. The team approach has become a logical consequence of these problems.

The question is sometimes asked, "What is so unique about craniofacial anomalies that they deserve to be singled out for special attention?"

The face of man is his window to the world. It contains the organs of sight, hearing and speech with which he communicates with his environment, receiving information and responding to it. The face reflects state of health, emotion and character. It is the facade by which others perceive and judge the individual. This most visible part of the body influences acceptability to society in general, and, more specifically, acceptability in human transactions. That which affects the face of man may also strike at
one of his most human functions—the ability to communicate through articulate speech.

The forming head is a community of interrelated cells, tissues and complex organ systems, varying considerably in developmental chronology. If in the temporal course of ontogeny, one member or constituent of this community is affected adversely, invariably other parts will suffer.

The head contains the portals for respiration, feeding and sensory perception. Structural or functional defects within the head of the newborn can be life-threatening in the neonatal period if aberrations in the oral-nasopharyngeal ports impede respiration or feeding. Arrest of growth of the neurocranium, if undetected and untreated during the period of rapid brain growth, can cause serious damage to the central nervous system.

The complexity of the clinical problems presented by such patients is not amenable to solution by a single discipline working in isolation from all other disciplines. Thus there is a need to facilitate communication between disciplines and to interphase their separate approaches to acquiring basic knowledge and to problem solving. To accomplish this, it becomes necessary to design a new mechanism for professional education in the health sciences that emphasizes the unity of the many problems presented by one patient.

This brings us to a discussion of the second element, the interdisciplinary team. There are different kinds of teams and each can be effective when matched to a specific mission. For example, to do open heart surgery one must have a multidisciplinary staff trained to function as an operational unit working largely under the command of a surgeon. In contrast, a different kind of team is required when one wishes to explore the no man's land that exists between the disciplines.

Our concern is with the team that sets out to remedy deficiencies in the existing pattern of professional education. It is a pattern that is departmentalized and treats knowledge in terms of components or fragments despite the fact that the life sciences are part of a continuum ranging from the molecular level at one end to the complexity of the total organism at the other. Collectively, the interdisciplinary team should have the conceptual and operational capability of entering this continuum at any level in order to solve a given problem. The interdisciplinary team possess an intellectual unity dedicated to the acquisition of knowledge. As a social unit it has succeeded in overcoming the impediments to communication that stem from barriers of jargon and the artificial jurisdictional boundaries created by the clinical specialties. Norbert Wiener described the utopian team:

"We had dreamed for years of an institution of independent scientists, work-
ing together in one of these backwoods of science, not as subordinates of some great executive officer, but joined by the desire, indeed by the spiritual necessity, to understand the region as a whole and to lend one another the strength of that understanding."

This brings us to the third element in the title and that has to do with the organization of the interdisciplinary unit. There was a time when federal funds were readily available and this seemed to spur every dental school and hospital to organize a cleft palate team. While the motives and intent may have been laudable, few survived as viable units and even fewer can be said to have contributed to the advancement of knowledge. As a result of this disappointing experience, what is the optimal setting for the interdisciplinary team?

A university health science complex with access to the varied resources of the scholarly community possess the required prerequisites. These prerequisites should include the behavioral sciences, speech and hearing sciences, bioengineering, biomathematics and computer science. In the clinical sciences, it must possess a core staff of full-time academics with a dedicated spirit of inquiry and a tradition of institutional commitment to research. Physical adjacency between collaborating departments and a tradition of intellectual companionability are additional ingredients that would augur well for the interdisciplinary unit.

What is the catalyst that provides the social and intellectual forces that bring diverse professionals into a collaborative egalitarian unit in which all are chiefs? It is a realization that the needs of certain patients cannot be resolved within the departmentalized structure of the hospital. It is a recognition that overspecialization bred cadres of specialists who knew more and more about less and less. It is a desire to counter this trend by making generalists out of specialists and in the process enhancing their value within their chosen field.

In the prevailing scheme the patient is placed on an assembly line where he is shunted from clinic to clinic, from one consultant to another, all the while fattening his hospital chart to the extent that the full record is seldom read, or if read, not understood. There is no organized mechanism for assessing all of the accumulated information, establishing priorities and sequences of treatment, or for evaluating the long-term results of therapy.

When a critical mass of clinicians becomes sufficiently concerned, it becomes possible to study the initial state of the patient in a systematic fashion, prescribe specific modalities of treatment, and determine the subsequent state through long-term follow-up. This effort can be sustained provided there is a commitment to a broadly based program of inquiry that is supportive to the educational needs of the staff. Coupled to this research
must be a recognition that assessment of therapy in terms of effectiveness and benefit to the patient as well as cost to society is a central responsibility of the interdisciplinary team.

Where such teams are effective, they can become a model for professional education and for the delivery of a health service that is truly dedicated to total patient care.

As such teams become established, they begin to attract an increasing variety of research talents. Such individuals are generally characterized as having pluralistic interests, as being possessed by the desire to comprehend the relation of their own interests to those of the broader regions of human biology.

Involvement with birth defects demands a wide range of knowledge that makes everything that one has learned relevant. It is a source of continued interest to observe middle-aged specialists, secure in their special sphere, study the new advances in molecular biology and seek instruction in clinical genetics as a result of the stimulus received within the interdisciplinary forum.

The impact of such teams on the academic community has transcended the application to their targeted mission with a measurable effect on the entire educational process. In colleges of dentistry this has led to a closer working relationship with speech scientists, geneticists, teratologists and other disciplines as students were attracted to the cleft palate team. This impact can readily be measured by the number and quality of papers submitted by dental investigators to the International Association of Dental Research, the American Cleft Palate Association and the Teratology Society, among other scientific societies.

Another by-product of the team’s involvement in problems of craniofacial biology has been a gradual dissolution of the unfortunate dichotomy that has separated the clinical investigator from the laboratory scientist. By tradition it seemed that the investigator lost the cloak of scientist once he left the laboratory to work in the ward, clinic or community. It was far easier to gain prestige through investigative work in the laboratory. As a cadre of clinical investigators trained in scientific discipline emerged from federally sponsored training programs, it became possible to build bridges between clinical and laboratory science. Clues derived from clinical experience had to be tested in the laboratory. What was needed was a clinician who could recognize the problem, formulate a hypothesis and suggest the means for testing it in the laboratory. Conversely, experiments which affect man cannot be duplicated in the laboratory at will and so afford the laboratory scientist a unique opportunity to study the interrelationships of abnormal form and function.
Lest the preceding seem idealistic and improbable in the real world to which we must return, consider the case history of cleft palate teams in this country. The story began with the enactment of the Social Security Act in 1935. Part II of Title V of this act provided for the establishment of Crippled Children’s Programs in all of the states and territories. While many states had such programs prior to 1935, the pattern was one which made medical care and related services available only to children with orthopedic difficulties. Thanks to the efforts of Herbert Cooper in Pennsylvania, these services were extended to include children whose problems were located anatomically in the head.

Pennsylvania became the birthplace for much of the early program in cleft palate. In 1943, Linwood Grace, D.D.S., chief of the Dental Division of the Department of Health, announced in the state dental journal that his department would sponsor a course in the construction of speech appliances for children with cleft palates, to be held at the Speech Clinic of the Pennsylvania State College. The course was taught by Cloyd S. Harkins, D.D.S., a prosthodontist, and Herbert Koepp-Baker, Ph.D., a speech pathologist. It was the first interdisciplinary course of its kind to be held in the United States.

As a result of that course, the American Academy of Cleft Palate Prosthesis was organized three months later at a meeting in Harrisburg, Pennsylvania, and Linwood Grace, D.D.S., was elected its first president. In 1949 the name of the organization was changed to the American Association for Cleft Palate Rehabilitation. This marked a change of emphasis from the purely prosthetic approach to include a broader interest in the person who had a cleft. In 1961, the organization was renamed The American Cleft Palate Association in an effort to minimize the emphasis on rehabilitation and reflect the larger concern that included prevention.

In retrospect, we can recognize a climate of political, social and scientific events that favored the development of cleft palate teams. First, there emerged the concept of treating the whole child. Secondly, this was augmented by the study and application of knowledge about growth and development. Thirdly, it was recognized that inadequacies arose from a system of rigid compartmentalization that was developing in specialty medical practice. And finally, interdisciplinary teams were increasingly utilized to deal with complex medical problems.

In the 1960's, federal support, largely through the National Institute of Dental Research, expanded the involvement of the scientific community not only in diagnosis and treatment but also in the search for etiologic factors and mechanisms by which clefts are produced, in the expectation that this would provide the means for prevention. This support provided the
basis for the comprehensive programs that are now flourishing at The University of Iowa and elsewhere.

Present events suggest that with the expansion of interest to include a wider range of craniofacial anomalies, the sophistication and scope of activities that now involve cleft palate teams will expand considerably, thereby increasing the opportunities for the dental scientist. This is already evident in the transition of some teams into centers for craniofacial anomalies or centers for communicative disorders.

Since no one institution encompasses the wide variety of talents required or possesses a sufficiently large catch basin of referrals to accrue an adequate case load of relatively rare entities, we are beginning to see a new type of team approach. It is a team involving a consortium of institutions. The arrangement maximizes the utilization of available resources and minimizes the cost.

Educators now recognize that there is a significant fallout from such enterprises that has a larger meaning to the curriculum. Increasingly, dental and medical students are being attracted to such centers. The dental student, long afflicted by a curriculum that overtrains for what he will do in private practice and undertrains for what he would like to do, finds that his education has not been wasted and that he can build on it when invited to participate in the team enterprise.

Of special interest was the reaction of medical students who intended to go into family practice. They find our conferences of value because we truly practice comprehensive medicine. Moreover, they have noticed that we are familiar with and make use of a variety of social agencies at the community and state level to assist in implementing patient care. They recognize the need to know about these resources if they are to be effective within a smaller community.

The interdisciplinary unit for the study of craniofacial anomalies has something for everyone. For the specialist, it makes him more of a generalist and enhances his rapport with other professionals, and as a result patient care improves. For the determined generalist, the center expands his horizons. The laboratory investigator derives important clues from the clinic that suggest critical experiments that may lead to advances in basic science.

In this larger enterprise dentistry has found a wide range for the exercise of its talents in dignified relation to other members of the health team.

References
FUTURE CARE OF THE HANDICAPPED PERSON

Richard Koch, M.D.

Introduction

Traditionally, the care of handicapped persons has been developed through the efforts of lay groups organized for the benefit of the physically or mentally disabled. While the care of physically handicapped children now has become largely a pediatric responsibility, this is still not true for the mentally handicapped in most of our larger states. In New York, Pennsylvania, Illinois, Texas and Ohio, the mentally handicapped child, be he mentally ill or retarded, is still primarily the responsibility of state agencies identified with the mental health professions of psychiatry, psychology and social work. In past years, this was true in large measure because the pediatrician seldom wished to become involved in the care of the mentally retarded in any measure beyond the usual procedures provided to any normal child. Today, however, services for the mentally retarded child in particular are changing rapidly. The spectacular advances in chromosomal technology and amino acid, lipid and enzyme biochemistry in the last few years have changed the care of the retarded child into an exciting chapter in the ongoing progress of medicine. Today we have the opportunity not only to correct the many physical handicaps of these patients, but to participate in prevention programs which will either eliminate the occurrence of many forms of mental handicaps, or at least minimize the degree of mental retardation present in affected individuals.

The problem of change

To accomplish a significant reduction in the prevalence of mental retarda-
tion will require change in our system of medical care. For it is in the health field that the significant reduction of mental retardation can now be accomplished. Change is unsettling. To some, it is considered radical; to others, it is considered progress. When change has an economic impact, you can rest assured it will be resisted by those affected adversely! There are many who say change cannot be legislated or imposed from above, but it is very clear that both legislation and the hierarchy of the health care system must together develop a climate that encourages change, so that those at the working level do have the opportunity to institute change, as opportunity to move forward develops. While most members of the medical profession have been fairly active in instituting change within their own profession, they have not been effective at the legislative level in their own respective states. If we are to move forward in the care of the handicapped in a constructive manner, we must develop more effective methods of encouraging legislative implementation of needed progress.

Changing from institutional custodial care to community based programs for the mentally retarded

In each of our respective states, most of the tax dollars spent for the mentally retarded are utilized to staff and maintain institutions for their care. Often these are located in rural settings where it is difficult to recruit staff and to institute training programs interacting with our university and college systems of higher education. In some states, the programs in these institutions are deplorable. In a recent visit at one of our large eastern states, I saw 120 to 160 residents in locked wards with absolutely nothing to do. They were occupying two large day rooms, in each of which four attendants attempted to keep order. There were no towels or soap at the wash basins and no paper in the toilets. Some patients were naked and lying on the floor in their own excrement. The odor was stifling. It seemed as if I had been transported back into the dark ages of despair. For this, the state was paying about $8,000 per patient per year for care!

Investigation reveals that the rate of construction of state institutions nationwide is still significant, in spite of the fact that professionals already know that the traditional custodial institution is not a part of the normalization principle that we so aptly espouse. Normalization refers to the process whereby mentally retarded persons are entitled to the same rights and privileges enjoyed by the normal populace. If a person becomes ill with cancer, he is not institutionalized in a locked ward somewhere out in the country! If we are truly committed to a principle such as normalization, then the first casualty in our present system will be the institution. Then why are we
building more of them? Could it be that we don't really believe in this principle, and that we are merely giving lip service to it?

The answer to this question is not very simple. In California, a decision was recently made by the state administration to close all state institutions for the retarded, and to work toward the development of community based services by 1982. Laudable as this goal was, the outcry from the public was, perhaps, predictable; however, it was astonishing and somewhat unanticipated by many professionals. But the announcement of the plans was abrupt and ill planned. There were inadequate high quality community based services to which the families involved could be referred. Thus the decision to work toward closure of the state institutions mobilized two very powerful groups which forced the state to retreat from the principle of normalization. They were the parents of retarded persons in the facilities and the California State Employees Association. The parents became very threatened by the idea that their retarded children no longer would have the "protection" of the institution, but would rather be "at the mercy of the community." The specter of "permanent parenthood" haunted them. After all, parents who were comfortable with their retarded child in a state institution would again become responsible for him if he were placed in a community setting. Many parents found this totally unacceptable. And can anyone really blame them? Having a retarded child is bad enough, but to have solved the problem by institutionalization, painful as that was, and now to be faced with the undoing of that decision, was simply intolerable. Many vicious letters were sent to state legislators who thought that they were doing the right thing in supporting the development of community services.

The role of the bureaucracy in self-perpetuation was also striking. In some states, employee associations have gone to court or threatened to strike when faced with alternates to the traditional methods of care in the state institutions. The important factor has been the preservation of state jobs and not what was best for the retarded person or his family. Crass as this may sound, it is absolutely true. There is something almost inherently evil in bigness. When organizations become so large that they lose sight of what is best for society, they begin to perpetuate the status quo to protect themselves. This is apparent throughout our society today. Witness the jurisdictional fight between the United Farm Workers and the Teamsters Union over farm labor! The dispute has disrupted agriculture, growers, workers and the consumer. For whose benefit? Certainly not for the consumer! Thus the normalization principle faces two very determined opponents. Any state adopting this principle must be prepared for such opposition.
The California experience in normalization

In 1965, California decided to implement the above policy by establishing community based services. In that year, Assemblyman Jerome Waldie introduced legislation authorizing the establishment of a Regional Center system funded by the state, but operated by private non-profit agencies such as parents associations, non-profit corporations and the like. The state legislature passed unanimously the bill (AB691) to establish this Regional Center system, and added a novel feature in that monies were added to the operating budgets of the centers that permitted them to purchase services for parents who were unable to implement center recommendations due to lack of funds. This unusual approach has been remarkable in its effect on the centers. To date, 15 centers have been authorized with a state budget of $22,000,000 for 1974. They are located throughout California so that no family lives over two hours' drive from a center. Each center offers multidisciplinary diagnosis, evaluation and counseling. The centers serve all ages and have been directed by the legislature to provide alternate solutions for parents whose retarded dependents require institutionalization, by purchasing private care whenever possible. There is no charge for diagnosis or counseling. If residential care is required, the family is assessed a monthly fee determined by its ability to pay, according to a sliding scale. Under no circumstances is a family required to pay more than it would to keep a normal child at home. All financial obligation ceases when the age of 18 years is reached by the retarded person. The effectiveness of the centers in reducing admissions to state institutions has been dramatic. In 1969, there were 270 admissions to Pacific State Hospital, whereas in 1972 there were only 39. The state institutional population has dropped significantly, from over 14,000 in 1965 to 10,000 in 1973. Similarly, the state had a waiting list for admission to its institutions of over 3,000 persons in 1965. Now there is no waiting list at all. In fact, the institutions today have vacancies. Therein, however, is a problem. As the number of residents in state institutions has declined, the staffing ratio has improved and now the programs in the institutions are improving markedly. Some years ago, the programs were primarily custodial. Today they are therapeutic. As a result, there will be increasing pressure upon the clinician to utilize a program in a state institution because it is better than a comparable community facility! It seems clear that the state must maintain model programs and provide leadership so that there will be constant improvement in all services. Ideally, it would seem as if a combination of state and community based programs is the answer to this complex problem of institution versus community. It seems
clear that, for the present, the retarded will be served best by a combination of these two models of care rather than by either alone.

The disappearance of the diagnostic clinic

Today we still have many diagnostic clinics, which do not follow up on patients and thus only begin the process of evaluation. As we have gained experience in providing multidisciplinary services, it has been realized that much professional time and effort is wasted due to lack of implementation of recommendations. Evaluation is a long-term process. Diagnosis can often be made in a one-time evaluation. Remediation, however, must be accomplished over a period of months or years, and without this effort, evaluation remains inconclusive. For example, one-fourth of the cases seen at the Child Development Clinic at Children's Hospital of Los Angeles, after treatment and follow-up were found to have been misdiagnosed or poorly evaluated. In a few cases the children were functioning at a lower level than was anticipated according to their diagnoses, but almost invariably, they were functioning at a higher level than had been predicted. Thus, more and more, there will be units in medical centers which will provide follow-up services to those persons who are diagnosed and evaluated. California now has 15 such centers in which no case is closed unless death occurs. Thus these units are providing services to an increasing age span. Pediatric age groupings, while characteristic of most diagnostic clinics today, will disappear tomorrow. This has import for our training programs. Another phase of these clinics is also disappearing. Until recently, there have been specialized clinics for mental retardation, cerebral palsy and epilepsy. These are now being combined under the designation "developmental disabilities." The broadening of responsibility is welcomed by most.

Advocacy for prevention programs

It is only recently that sufficient technology has been available to institute significant effort to prevent the birth of mentally retarded individuals. Throughout our country today, there are some exemplary programs such as the care of the premature in certain states or the Massachusetts Metabolic Screening Program. But these are few. Why is it that even today, our population is not totally protected against measles, poliomyelitis, diphtheria, tetanus, pertussis and German measles? Adequate amounts of vaccine are available, and certainly manpower is sufficient. It would almost seem as if our priority for national effort is greater in the bombing of Southeast Asia than in the prevention of birth defects right here at home. Is it the system of providing medical care that is at fault? Or is it the inherent lethargy of people in making changes? Perhaps the lay public is largely unaware of the
untold tragedies which result from unnecessary illness. However, it seems inexcusable that the medical profession has not done more to educate the public to the importance of preventive measures. If the public knew today that 25 to 50 percent of the births of mentally retarded children could be prevented, it would demand action. It is now well known that babies whose mothers have had German measles during the first trimester of the pregnancy are nearly always born with multiple anomalies, including mental retardation; however, it is not well known that there is now a vaccine which can give lifetime protection against this disease. It is no longer necessary for any woman to run the risk of contracting German measles during her pregnancy, however little has been done to publicize this important development. On the contrary, the Medical Society in California, on the advice of the Public Health Service, has been adamant in refusing to administer this vaccine to any girl over the age of 13 years. The reason for this seemingly reprehensible decision is the fear on the part of most doctors of malpractice suits. Their reasoning is that the vaccine might cause damage to the fetus if it is administered to a pregnant woman, and any girl 13 years old has probably reached puberty and therefore might be pregnant. There is at least one case in which a woman who was denied the vaccine, subsequently became pregnant, contracted German measles during the pregnancy, and gave birth to an abnormal child. She has brought suit for malpractice against the doctor who refused her the inoculation. This case may provide a precedent so that doctors will take a more realistic approach to this problem in the future. *Every girl who has not had German measles by the age of 12 should be given the vaccine.*

It seems to me that when those of us who are aware of the facts are too fearful to act, we are facing an almost insurmountable job in our efforts to rally public opinion to our cause. Noble as the efforts of some may be to bring about change in the system, with the present economic strength inherent in the various professions and hospitals change will be virtually impossible to achieve. But change must occur, and will occur. An aroused citizenry will not allow our present acute-crisis-oriented form of medical care to function in its present method much longer. Thus it is incumbent upon us to lead the way so that change will not be disruptive. What are some of these changes that will effect us in the near future?

Better maternity care is mandatory. The number of abortions has had one unexpected benefit: obstetricians are less busy, and have had more time to think about how to provide better services to the populace as a whole. The changes in our welfare system allowing the indigent to utilize community services and private practice facilities have been a significant step forward. The development of the nurse-practitioner in prenatal care is another im-
portant aspect of improving maternity care. But these are not enough. More must be taught in our public schools about the need for good prenatal care. Our citizens must be taught that the premature birth rate is related to economic factors. How ridiculous of society to be willing to pay for 30 years’ care in an institution at $10,000 a year for a retarded person, but unwilling to pay for the small cost of good prenatal care! The rate of prematurity in poverty areas is four to five times that in our well-to-do suburban areas. Since prematurity is directly related to the problem of mental retardation, we must make every effort to reduce it to the lowest possible figure.

Better maternity care would reduce the occurrence of toxemia of pregnancy, placenta praevia, traumatic birth, etc. The prospective mother should know that cigaret smoking predisposes her babies to a smaller birth weight. She should know of the great danger of maternal German measles to her unborn child. Mothers with diabetes and diseases such as phenylketonuria should be counseled against having babies of their own, since the occurrence of congenital anomalies is greater in a mother with any chronic disease. She should know of the importance of being inoculated against the Rh antibody in situations of maternal Rh negativity. In short, the tools are now available to prevent the birth of many types of handicapped persons.

Perhaps the public schools should share in the responsibility for bringing about the necessary education of the students so that they will demand more progressive medical care when they become adults. The sex education and health classes could perhaps be combined into family relations classes, which could encompass not only sex education and health, but preventive medicine and drug education. Such a program might take away the stigma of the title sex education, which has become a scare phrase to many people, and the students could be provided with a well-rounded fund of information which could be invaluable to them in later life.

Screening newborn populations

Some years ago, routine screening of newborns became a reality for phenylketonuria. While pediatricians for years have been advocating routine physical examinations for newborns, they have felt uncomfortable about routine biochemical screening. In fact, one famous venerable pediatrician, when asked about routine biochemical screening, snorted his disgust at this idea by saying to his students, “We are trying to teach you to be clinicians and not to be dependent upon any laboratory test.” Noble as this concept might have been at one time, modern pediatrics will more and more include routine screening for a variety of diseases. Twenty years ago, the blood count, tuberculin skin test and a test for syphilis were the only routine lab-
oratory studies performed, and they were only done on patients requiring hospitalization. In the near future, all children will be screened for a variety of metabolic diseases as well as chromosomal and other genetic conditions. Technology hopefully soon will allow us to identify hypothyroid function before mental retardation occurs. At Pacific State Hospital, one percent of the residents are there because of hypothyroidism. Good genetic counseling services will eliminate or significantly reduce such disorders as Tay-Sachs disease, Hurler's syndrome, Niemann-Pick's disease, Gaucher's disease, etc. Amniocentesis will become a household word, and be performed on all pregnant women over 35 years of age. The fluid will be utilized for not only chromosomal disorders, but for routine enzyme studies, metabolic evaluation and for viral culture as well.

Abortion, birth control, sterilization and sexuality

The abysmal ignorance that exists today in our populace about normal reproduction is a sad commentary about our modern society. It suggests that our technology has greatly outstripped acceptance of many of our social values.

Today the topic of sexuality can be discussed in magazines, newspapers, radio, movies and television. Progress is occurring. The advent of mass media will do much to create a climate of change in attitudes. The Supreme Court decision regarding abortion has finally obtained an important goal for those of us interested in preventing the birth of severely malformed persons. Without this decision, the term genetic counseling had a hollow echo. Abortion should be the responsibility of the family involved and their physician, and should not be cloaked in religiosity or psychiatric jargon. It should be freely available regardless of economic circumstances. Birth control, sterilization and sexuality have become more or less accepted for the normal population, but not as yet for the handicapped. Sterilization for them is still a complicated legal problem. We have not resolved our feelings about marriage for the mentally retarded. While marriage is a step toward normalization for the retarded person, there are serious reservations for most that the right to have children of their own should remain restricted.

Conclusion

Change is upon us. It is our responsibility to make these changes constructively with the help of an aroused citizenry that will be demanding much of us. I am confident that we can meet this demand.