Second Edition

NEUROLOGICAL SURGERY

Volume 3

A Comprehensive Reference Guide to the Diagnosis and Management of Neurosurgical Problems

Edited by

JULIAN R. YOUMANS, M.D., Ph.D.

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1982 W. B. SAUNDERS COMPANY
Philadelphia • London • Toronto • Mexico City • Sydney • Tokyo

W. B. Saunders Company: West Washington Square
Philadelphia, PA 19105

1 St. Anne's Road
Eastbourne, East Sussex BN 21 3UN, England

1 Goldthorne Avenue
Toronto, Ontario M8Z 5T9, Canada

Cedro 512
Mexico 4, D.F. Mexico

9 Waltham Street
Artarmon, N.S.W. 2064, Australia

Ichibancho, Central Bldg., 22-I
Chiyoda-ku, Tokyo 102, Japan

Library of Congress Cataloging in Publication Data

Youmans, Julian Ray, 1928–
Neurological surgery.

1. Nervous system—Surgery. I. Title.
[DNLM: 1. Neurosurgery. WL368 N4945]
RD593.Y68 1980 617'.48
ISBN 0-7216-9664-3 (v. 3) 80-21368

Volume 1 ISBN 0-7216-9662-7
Volume 2 ISBN 0-7216-9663-5
Volume 3 ISBN 0-7216-9664-3
Volume 4 ISBN 0-7216-9665-1
Volume 5 ISBN 0-7216-9666-X
Volume 6 ISBN 0-7216-9667-8
Six Volume Set ISBN 0-7216-9658-9

Neurological Surgery—Volume Three

Last digit is the print number: 9 8 7 6 5 4 3 2 1

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DEVELOPMENTAL AND ACQUIRED ANOMALIES

GENETIC ASPECTS OF NEUROSURGICAL PROBLEMS

A "birth defect" can be defined as a condition resulting from an insult to the embryo during pregnancy or from transmission of a hereditary trait. This broad definition includes conditions such as those resulting from the effects of teratogenic agents or secondary to chromosomal aberrations. The bulk of genetically transmissible disorders is due to single gene defects, and of 2336 listed in McKusick's *Mendelian Inheritance in Man*, a third are of importance in the field of neurological genetics.[71]

Neurological genetics embraces both delineation and clinical management of birth defects affecting the nervous system. It elucidates the clinical picture and provides knowledge of potential complications necessary for proper management. It also seeks to define the risk of recurrence of a defect and to provide the family or disabled person with genetic counseling. Neurological surgeons can complement medical management by operative correction of a birth defect, and this can lead to fruitful collaboration between neurosurgeons and neurogeneticists. In this chapter, birth defects are described with emphasis on problems that neurosurgeons may encounter. Only a brief description of basic genetic principles is given.

NEUROLOGICAL GENETICS

Basic Mendelian Traits

The usual assumption in mendelian genetics is that the trait to be studied is under the control of a single genetic "locus" and obeys the laws of mendelian inheritance. The assumption is sufficient for applying mendelian rules derived from study of large populations to particular clinical situations. Mendel described transmission of traits from one generation to another and defined them as "dominant" or "recessive" on the basis of their modes of inheritance. In Mendel's words,

> Those characters which are transmitted entirely, or almost unchanged in the hybridization, and therefore in themselves constitute the characters of the hybrid, are termed dominant; and those which become latent in the process, recessive.[71]

Mendel also developed "rules" by which the geneticist can predict, or at least expect, the transmission of known characteristics from parents on through succeeding generations. Although it is not the purpose of this chapter to provide extensive detail concerning the transmission of heritable characteristics, a few basic points permit ready understanding of mendelian principles and their application to populations, families, and individuals.

In meiosis, paired parental genes are segregated so that each gene has a 50 per cent chance of being transmitted to the germ cell. Each offspring normally receives a single set of genes from one parent and a complementary set from the other parent. If one or both genes of the pair are dominant, the trait governed will be expressed to at least some degree in the offspring. If a recessive gene is paired with a dominant gene the recessive gene normally will be "masked" by

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the dominant gene. Only when both of the paired genes are recessive will the recessive trait be expressed in the offspring. As applied to genetics, the terms "dominant" and "recessive" refer only to the interactions among genes at the same locus and their expression in the phenotype. The terms carry no implication of fitness or lethality. The transmission of a character can further be related to certain chromosomes, the class of autosomal chromosomes and the X-chromosome.

Autosomal Dominant Transmission

An autosomal dominant trait is suggested when a particular phenotype is expressed through several successive generations, affects both males and females, and is transmitted from either of the parents and notably from father to son. The segregation of the autosomal dominant trait is also independent of the sex of the offspring and follows the classic rule of 50 per cent, assuming only one of the particular parents carries the dominant trait. When both parents carry the same dominant trait, 25 per cent of their children will receive two dominant genes for the trait. If the dominant trait in such instances is deleterious, the phenotype will be incompatible with life in the homozygote (e.g., achondroplasia, von Recklinghausen disease, Huntington chorea).

Autosomal Recessive Transmission

In autosomal recessive segregation, phenotypically normal parents may produce a rare phenotype in a sibship. Consanguinity of parents and geographical or cultural isolation are factors that increase the incidence of recessive traits, especially the very rare ones. The usual situation arises from mating between phenotypically unaffected parents (carriers), which results in 25 per cent of descendants of both sexes being affected. Recessive disorders usually manifest themselves earlier in life than do dominant conditions, and in several instances the recessive conditions are genetically lethal. Genetic lethality has no relation to the longevity of the individual. It means that as a result of the inherited condition an affected individual does not have descendants. It must be stressed that carriers for a strictly autosomal recessive trait are phenotypically normal. Detection of such heterozygotes by special tests is the rationale for screening programs, such as that for Tay-Sachs disease.

X-Linked Transmission

At present no genetic pathological trait is known to be transmitted on the Y chromosome, and therefore the designation of "X-linked" is more appropriate terminology than "sex-linked." Because of having but one X chromosome, the male phenotype for any trait carried on this chromosome corresponds exactly to his genotype. The critical element by which X-linked heredity (recessive or dominant) is recognized is the absence of father-to-son transmission. In X-linked recessive heredity (e.g., hemophilia, Fabry disease) all sons of an affected male are phenotypically and genotypically normal. All daughters of such a father are phenotypically normal but carriers of the trait. On the average, a female carrier of the X-linked trait will transmit the disorder to half her sons. Also on average, one of two of her daughters will carry the trait, the others having a normal genotype. Situations that are difficult to interpret may occur if an affected male and a female carrier have children.

X-linked transmission differs from sex-limited heredity. In sex-limited inheritance, the condition occurs only in one sex, whereas in X-linked heredity, the transmission is through the X chromosome, and the trait may appear in both males and females.

Variability and Heterogeneity

A major concern in medical genetics is to delineate the relationship between a given phenotype and an already identified clinical entity. The same genetic defect may produce different phenotypes (variable expression, or variability), or identical phenotypes may be produced by different genes (genetic heterogeneity).

Variability exists because a gene operates in combination with the environment and the genetic background of the individual. It is most evident in conditions exhibiting marked pleiotropy or multiplicity of effects.

Heterogeneity exists when segregation occurs according to two different transmis-

sion models, as with Hurler (recessive) and Hunter (X-linked) diseases. The present inability to distinguish clinically between different phenotypes does not contradict the principle of heterogeneity. Charcot-Marie-Tooth disease is certainly a heterogeneous condition, as shown by the existence of dominant, recessive, and X-linked forms. In very general terms, if several transmission models are known, the clinical picture is mildest in the dominant form, most severe in the recessive model, and intermediate in its effects in the X-linked form. Another clinical clue that suggests heterogeneity is variation in the age of onset and mode of evolution of the disease. Werdnig-Hoffmann disease was thought to be homogeneous until it appeared that some patients became disabled in their teen-age years and had a protracted course of evolution.

Genetic Counseling

Valuable advice can be given to concerned parents regarding their chances of producing a child with a disability and the possibilities for its treatment. Numerous advances in surgical and medical techniques offer affected individuals and their families options that hold promise of a normal or nearly normal life despite conditions that once were beyond the skills of the practitioner.

Applications of genetic information that would be impossible, if not inhumane, when applied to populations are an entirely different matter when applied at the level of the individual family to prevent the burden of a severe phenotype. Genetic counseling provides the family with accurate risk figures and, for individuals, defines the chance of manifesting a given condition. Furthermore, greater awareness of the potentials on the part of individuals, families, and physicians will also improve the likelihood of earlier detection and management of disorders.

Geneticists use three types of information to arrive at decisions: empirical, modular, and particular. Empirical information is that accumulated through experience—mutation rates, gene frequencies, penetrance coefficients, and their relationships to age, sex, and age at onset of the disease. Empirical data derive from statistics collected from a given population at a given period of time. If used with caution, the empirical approach can provide a definite basis for modifying risk estimates for the occurrence of a condition.

Modular information is knowledge of the mode of inheritance of the condition derived from genetic analyses and from the nature of the condition. For example, it has been observed that dominant conditions tend to be disorders of structure, having relatively late onset, and are mild enough to permit transmission of the trait; and recessive conditions are more likely disorders of function with earlier onset, tending to be more disabling.

The third source of information is that "particular" to the person seeking advice, who is termed the "consultand." Information from this source derives from the family pedigree, including history of similarly disabled individuals in the particular family.

A basic step in counseling is to determine whether the genotypes of the parents are known. If the genotypes are known, and assuming complete penetrance of the gene, the classic percentages of mendelian transmission can be invoked. If the genotypes of the parents are unknown, then computation of the probability for transmission of a trait is more complex. The method for estimating probabilities is of little interest in itself. Some authors prefer a precise estimate of the probability; others are satisfied with a quotation of "high," "low," or "no" risk. Parents of an affected child take little note of the mathematics. What concerns them is the combination of the risk and the burden of the genetic condition. Under the term, "burden," Murphy introduced the notion of the "price" of the genetic condition.[77] The price of a birth defect is not merely the financial cost of treatment; it also includes the emotional investment over the span of the condition. The burden can be intense and of short duration, as in the birth of an anencephalic child. Or it can be long-standing and of small significance, as with color blindness. Curiously enough, the concept of the burden of birth defects has seemed too trivial for exploration except by a few geneticists, whereas it is by far the greatest concern of parents. A distinction should be made as well between the burden for parents and the burden for the disabled individual.

Referral to a neurogenetic unit should take into account the limitation in human nature on the amount of "bad news" that can be absorbed at one time. A three-stage approach is more likely to succeed. In the first stage, a referral to a neurogenetic unit will help delineate the problem and the possibilities of operative treatment. In the second, correction of the birth defect can be accomplished. In the third, a referral to a neurogenetic unit for medical follow-up and genetic counseling will be rewarding even though, knowing that operative management is under way, parents are likely to be unable or unwilling to accept counseling on the risk potential for successive children.

As with other aspects of medical and surgical care, attention should be given to legal implications, including appropriate referral, management, and informed consent. Von Hippel-Lindau disease is a good example of a condition in which failure to refer a family for follow-up and genetic counseling may have serious consequences. With this condition, proper evaluation and, eventually, neurosurgical treatment can be helpful for the patient and for 50 per cent of the cases in his family.

CONGENITAL MALFORMATIONS

Epidemiology of Congenital Malformations

Estimates of the incidence of birth defects vary widely, depending upon the definition of the term, the sources of the sample, and the age of the groups studied. Among stillborn infants, the incidence may reach 31.5 per cent; it may be 17.8 per cent in those who die during the first week of life.[80] Among surviving newborn children the incidence is estimated to be 5.4 to 7.4 per cent.[110] Major congenital anomalies show a frequency of 2 to 3.3 per cent, and minor variants and structural abnormalities[31] one of up to 9.6 per cent. The usual figure quoted to parents for the risk of birth of a congenitally malformed child is 2 per cent. Other factors such as the age of the parents and the family history should also be considered when giving a recurrence risk figure for congenital malformation.

Central nervous system anomalies are underreported. Malformations such as polydactyly do not escape attention, but minor central nervous system malformations do despite their much greater implications for the useful life of the child. From the data in the British Columbia register of handicapped children it is possible to compute the incidence of some birth defects.[81] Strabismus and club foot are the leading diagnoses; however, spina bifida, meningocele, and hydrocephalus are frequent also (Table 34–1). In a World Health Organization study of 24 centers in which there were 417,000 single births during the years 1961 to 1967, the incidence of central nervous system anomalies was 2.66 per 1000 births.[109] The most frequent malformations were anencephaly (0.92), hydrocephalus (0.61), and spina bifida with or without meningocele (0.55). Central nervous system malformations are cited infrequently as cause of death. Of 1196 death certificates listing congenital malformation as one of the causes of death, only 6.7 per cent mention a malformation of the central nervous system as the primary cause. There are also variations in the geographic distributions of central nervous system defects that are reported. The lowest rate was reported by Japan (0.5 average annual age-adjusted death rate per 100,000 population) and the highest by Ireland (6.8 per 100,000 population). The average of incidence rates reported by most countries participating in the World Health Organization study was within the range of 3.0 malformations per 100,000 population.[95] Over the past 20 years there has been a steady decrease in the number of deaths attributed to central nervous system birth conditions. In the United States the rate declined from 24.1

TABLE 34–1 INCIDENCE OF SOME BIRTH DEFECTS*

CONDITION	PER 1000 BIRTHS
Strabismus	2.30
Club foot	2.29
Cardiac malformation	1.70
Cleft palate and harelip	1.70
"Cerebral palsy"	1.40
Mongolism	1.00
Spina bifida and meningocele	0.94
Hydrocephalus	0.59

* After Newcombe, H. B.: Population genetics: Population records. *In* Burdette, W. J., ed.: Methodology in Human Genetics. San Francisco, Holden-Day, Inc., 1962, pp. 92–113. Data from the British Columbia Register of Handicapped Children.

TABLE 34–2 APPROXIMATE TIMING OF VARIOUS MALFORMATIONS DURING HUMAN MORPHOGENESIS

MOMENT OF INJURY	TYPE OF MALFORMATION	EXAMPLE
Blastogenesis	Defective separation of twins	Craniopagus twins
Embryogenesis		
24th day	Defects of neural tube closure	Anencephaly
26th day		Meningomyelocele
5th week	Anterior midline defects	Arhinencephaly
5th week	Failure in development of pontine flexure	Arnold-Chiari malformation
6th week	Defective development of rostral membraneous area	Dandy-Walker syndrome
6th week	First visceral arch syndromes	Craniofacial anomalies
7th week		Cleft palate
Fetogenesis		
11 to 13th week	Defects in neuronal migration and layering	Agyria
13th week		Pachygyria
16th week		Microgyria
20th week		Neuronal periventricular heterotopias
5th to 8th month	Defects in commissuration	Agenesis of corpus callosum

per 1000 deaths in 1950 to 17.8 per 1000 deaths in 1967, with major declines noted for spina bifida and hydrocephalus. Such declines parallel the decrease in national live birth rates during the same period, and more data are needed to ascertain whether there has been any impact of better neurosurgical management.

Various teratological and embryological studies have shown a relationship between the period of injury to the embryo and some types of malformations (Table 34–2). An injury during the early stages of blastogenesis may induce defective separation of twins and result in malformations such as craniopagus twins. Spinal dysraphias are engendered during the third to fourth week of embryogenesis. Defects in neuronal maturation and failures in commissuration may result from insults during the last trimester of pregnancy. Warkany's *Congenital Malformations*, Bergsma's *Birth Defects. Atlas and Compendium*, and Smith's *Recognizable Patterns of Human Malformation* are comprehensive reference textbooks that may be consulted for further discussion of these matters.[8,106,120]

Craniopagus Twins

Craniopagus twins are an object of curiosity to laymen and physicians alike. From various reports it appears that the site of the junction is the most important criterion for successful operative treatment. The site can be frontal, parietal, or occipital. Concomitantly there can be rotation of the heads in the coronal plane and various degrees of angulation between the longitudinal axes of twins' bodies. The coronal plane rotation can result in the heads' facing in the same or opposite directions. The angulation in longitudinal axis may result in alignment of the twins' bodies as in vertex parietal junction, or in formation of an acute angle in lower frontal or parietal junctions. The interface of the junction is most important, being relatively small in frontal junctions and larger in parietal junctions.

Problems encountered during operative procedures are related to four aspects of the malformation.

Plane of Cleavage

In only 1 case of 14 was there a thin sheet of bone between the heads.[117] In frontal craniopagus twins, a dural leaf delineating a plane of cleavage is likely to be present. In more posterior junctions, a dural shelf may be present on one side of the junction, a leptomeningeal coverage on the other. Only leptomeningeal separation can be expected in most cases, but it can be very dense, as described by Baldwin and Dekaban.[4]

Venous Connections

Common sagittal sinus is frequent in parietal and occipital junctions. Other venous connections are numerous. The dural shelf may contain the sagittal sinus, and the survival of one of the twins may depend on to which the sinus is allocated.