EDITORIAL

Congenital Complete Heart Block

SEVENTY years ago Morquio described a family in which several siblings had slow pulse with syncopal episodes and death in childhood, and first proposed the concept that complete heart block may be of congenital origin. Since then abundant case reports in the literature attest that this entity, uncommon but not rare, has provoked great curiosity and has been documented to occur with a wide range of clinical and necropsy findings. Histopathological observations of the conduction system in congenital complete block has been extended by Lev et al. with the publication in this issue of Circulation of two cases which indicate disruption or discontinuity of the atrioventricular (A-V) junctional tissue and bundle branches. Electrophysiological studies with His bundle catheter recording techniques promise further understanding of the sites of ventricular pacemaker origin and more precise functional and pathological correlations.

The earliest clue to the presence of congenital heart block may be the detection of bradycardia in utero, often misinterpreted as an obstetrical sign of fetal distress. Electrocardiograms at birth or at the time of initial diagnosis usually show complete A-V block with ventricular rates in the 40's and atrial rates appropriate for the age. Most infants with complete heart block as their sole cardiac abnormality grow and develop normally. Children and adults are usually asymptomatic and lead active lives; many have participated in competitive sports, and some have undergone normal pregnancy and delivery.

The two major factors influencing symptomatology and prognosis are: (1) the site and stability of the ventricular pacemaker and (2) the presence or absence of associated congenital heart disease. These are of primary importance in anticipating the problems of, respectively, Stokes-Adams attacks or congestive heart failure. Such severe complications, or even sudden death, are particularly prone to occur in infancy or early childhood.

The ventricular pacemaker in congenital complete heart block, with or without cardiovascular malformations, is usually a single and stable focus originating in A-V junctional or His bundle tissue. It is characterized electrocardiographically by narrow, normal-appearing QRS complexes of uniform contour with R-R intervals that are usually constant; ventricular rates at rest are most frequently in the range of 40–55 beats/min and may increase slightly with exercise. Occasionally, the QRS duration for the age may be borderline to definitely prolonged (0.10–0.14 sec) with a configuration resembling left, or less commonly right, bundle-branch block; ventricular rates in this group as a whole may be a little slower and in the range of 35–45 beats/min. Ventricular rates at birth under 30 beats/min are often associated with multifocal QRS complexes, and suggest prenatal inflammatory or toxic processes affecting the conduction system, rather than a developmental
defect. An additional or ectopic ventricular focus, which may appear even in the context of a predominantly regular ventricular bradycardia with unifocal complexes, may be of ominous import. It may predispose to instability or shifting of ventricular foci which, rather than absolute rate, appears to be the critical factor in provoking dangerous ventricular tachyarrhythmias or Stokes-Adams episodes. Among the altered conduction patterns reported to occur in individuals with congenital complete heart block has been that of anomalous A-V excitation of the Wolff-Parkinson-White type.6

Congenital heart disease frequently coexists with congenital complete heart block, but its precise incidence is difficult to determine. Estimates vary according to (1) the extent of documentation by cardiovascular laboratory studies of the origin of murmurs and underlying anatomy and (2) the age of the population surveyed. Isolated ventricular septal defect, once thought to be almost invariably a concomitant lesion with congenital heart block, has been shown by cardiac catheterization to be an infrequent finding. Approximately one-half of the children with complete A-V block followed in a cardiac clinic setting are noted, however, to have some associated cardiovascular malformation. The incidence of malformations, especially severe ones, may appear higher in infancy because the signs and symptoms attract early clinical attention and many of the patients succumb before childhood.

Though an infinite variety of simple and complex cardiovascular malformations may accompany complete heart block, the most important statistical association is with the form of transposition complex referred to as "corrected" or "L-transposition" of the great vessels, the diagnosis of which may be readily established by angiocardiography.6,10,11 The designation refers to the unusual embryologic development of a levo or leftward bending of the primitive cardiac tube (as opposed to the usual D, dextro or rightward loop) in which the morphology of the ventricles is inverted, i.e., the right-sided ventricle has the structure of a left ventricle. Normal circulatory pathways and hemodynamics are not altered by ventricular inversion with transposition of the great vessels unless, as is common, intracardiac malformations, e.g., ventricular septal defect, exist. Complete heart block may occur with corrected transposition when the latter is an isolated morphologic pattern or is associated with other defects. It is of special interest to note that whereas heart block is usually recognized in infancy, conduction defects with corrected transposition may not be detected until childhood or adult life; partial forms of A-V block as well as complete block are common.

Congestive heart failure with complete heart block seldom occurs except in infants with underlying heart disease. Slow, regular ventricular rates per se probably do not cause cardiac decompensation though they may be a contributing factor.6 The structural cardiovascular malformations that are a particular liability include the relatively common ones, e.g., patent ductus arteriosus and coarctation of the aorta, which because of their size or severity may predispose to the development of congestive failure in the neonatal period. Intensive cardiotonic treatment should be pursued and surgical repair carried out whenever possible to relieve the hemodynamic stress imposed by the associated cardiac defect.

While complete heart block of congenital origin is generally a stable condition with good prognosis for survival, heart block following surgical repair of intracardiac malformations, such as ventricular septal defect, tetralogy of Fallot, and endocardial cushion defect, is a serious, though now infrequent, complication that may cause sudden death.12 Thus, permanent cardiac pacemakers are almost always implanted for persistent postsurgical complete A-V block, whereas they are rarely needed in congenital block.

Artificial pacemakers have been successfully utilized on occasion in congenital complete heart block for severely symptomatic infants or children who have manifested either (1) Stokes-Adams syncope with ventricular dys-
rhythmias or (2) unremitting congestive failure with associated congenital heart disease. Temporary cardiac pacing by means of a transvenously placed catheter electrode may be helpful in controlling ventricular rhythm or in assessing the clinical response to acceleration of ventricular rate, but it is not satisfactory as a method of chronic pacing in infancy. For permanent pacing, thoracotomy is presently required for implantation of epicardial electrodes. A variety of lead systems and pulse generators similar to those used in adults has been employed, including fixed-rate and ventricular demand pacers and atrial P-wave synchronized units; radio-frequency pacemakers have been established in a few patients with congenital block. Once a pacemaker is implanted, commitment is made for prolonged or, most likely, life-time pacemaker dependence. The ever present threat of pacing dysfunction or failure requires the availability of experienced medical and surgical personnel; complications are frequent, and multiple operative interventions are almost always necessary. Since adaptation to the currently available models of implanted pacemakers is a formidable course for the child and his family, the indications for their use should be very clear. It is to be hoped that future advances in pacemaker technology will lighten the burden and hazards attendant on the care of infants and children with these devices.

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