Prognosis for Patients with Congenital Heart Disease and Postoperative Intraventricular Conduction Defects

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SUMMARY Intraventricular conduction defects are common following repair of various forms of congenital heart disease. Such defects may affect adversely the long-term prognosis of patients in whom cardiac hemodynamics were adequately restored. Review of previously published studies suggests that the site of the conduction defect may be the reason for the different prognoses reported for patients from different institutions. The so-called "trifascicular block" pattern which sometimes occurs following open heart surgery is probably due to a more extensive lesion to the branching and penetrating parts of the His bundle rather than additional injury to the posterior left bundle branch fibers. Transient complete heart block in the immediate postoperative period seems to be a predictor for late development of complete heart block or sudden death at least as powerful as right bundle branch block and left anterior hemiblock.

CONSIDERABLE CONCERN has been expressed in recent years regarding the long-term prognosis of patients who develop various types of ventricular conduction defects following open heart surgery.1-11 Such intraventricular conduction disturbances occur in most patients with various forms of congenital heart disease following correction of their intraventricular defects through a right ventriculotomy,1, 2, 15 resection of intraventricular muscle containing parts of the specialized conduction system19 or due to septal sutures used for correction of the intraventricular defects.5-7, 13, 14

In most patients such conduction defects appear immediately upon termination of surgery and persist in almost all cases throughout their lifetime. Late development16, 18, 19 and late disappearance20 of various types of intraventricular conduction defects have also been described.

The desire of the medical community to understand the nature of such defects, to identify the location of injury, its electrophysiologic effects and electrocardiographic sequelae and to predict with precision the effect on long-term prognosis is therefore easily understood. In spite of the various pathologic11, 4, 17 and clinical studies,2, 4, 8, 9 significant controversy still exists regarding the precise location of injury in many of these patients, the ability to identify the exact location by electrocardiographic techniques and the long-term prognosis for such patients.

Right Bundle Branch Block Pattern

Right bundle branch block pattern is the most common intraventricular conduction defect following open heart surgery in the pediatric population. It has been reported to occur in from 60 to almost 100% following repair of tetralogy of Fallot,1, 2, 11 in 60 to 81% of patients with ventricular septal defect repaired via a ventriculotomy,21, 22 and in almost 100% of patients following repair of complete A-V canal.23

A right bundle branch block pattern on the electrocardiogram also occurs in 25 to 44% of patients with ventricular septal defect repaired via the atrium without a right ventriculotomy,21, 22 following repair of ostium primum defects29 and following correction of intraventricular communications in virtually all other forms of congenital heart disease.

In most patients who had a right ventriculotomy for repair of their intraventricular defects, the right bundle branch block developed because of peripheral disruption of the right bundle branch when the right ventriculotomy was carried out.12, 13 In others, proximal interruption occurred when septal sutures were placed to close the interventricular septal defect,5, 7, 14 and in still others, following isolated pulmonary infundibular resection.13, 24 In short, the right bundle branch may be injured along its entire course when open heart surgery is carried out in its proximity.

The precise location of injury to the right bundle branch can be ascertained in patients in whom only a single surgical procedure such as septal sutures or infundibular resection has been carried out. However, the identification of the precise location of injury is more complex in cases in which several procedures that may cause a right bundle branch block were carried out, such as in patients with tetralogy of Fallot who had a right ventriculotomy and ventricular septal defect closure and infundibular resection. In such patients injury to the right bundle branch may occur as an isolated or combined lesion with each of the surgical procedures.5, 13

The effect of the surgically created right bundle branch block pattern on long-term prognosis is unknown. It can be assumed reasonably that prognosis is related to the location of injury. The more distal the site of injury, the higher is the likelihood for intact right bundle branch fibers to exist and allow normal atrioventricular conduction even in the presence of a complete left bundle branch block. On the other hand, the more proximal the lesion, the higher the likelihood for such patients to develop late complete heart block upon later development of a left bundle branch block. This eventuality has been described recently in a patient who developed complete heart block upon subsequent development of injury to the left bundle branch.4

Since long-term prognosis is dependent upon the site of injury to the right bundle branch, it can be assumed reason-
ably that two populations of patients with postoperative right bundle branch block will be detected in the future: the first with peripheral right bundle branch block in whom prognosis probably will not be significantly affected, and the second with proximal right bundle branch lesions in whom the likelihood for complete heart block in later life will be higher.

Right Bundle Branch Block and Left Anterior Hemiblock Pattern ("Bifascicular Block")

The development of right bundle branch block pattern on the electrocardiogram associated with a counterclockwise and superiorly oriented frontal QRS complex (the so-called "left anterior hemiblock" pattern) is less common following open heart surgery and develops in 8 to 22% of patients operated upon for tetralogy of Fallot. It also occurs following repair of membranous septal defects either through a ventriculotomy or via the atrium.

The reported prognosis for patients who develop postoperative bundle branch block and left anterior hemiblock pattern on the electrocardiogram vary considerably. Increased incidence of late development of complete heart block and sudden death has been reported from some centers whereas no increase in these late complications have been reported by others.

It seems that the disparity in the reported clinical observations and prognosis can be explained by different mechanisms for this identical electrocardiographic pattern and by different sites of injury to the specialized conduction system which have occurred intraoperatively in these two different patient populations. As previously noted, the right bundle branch may be injured by various surgical procedures along its entire length. In addition, left bundle branch fibers oriented toward the anterior part of the left ventricle may also be injured during closure of isolated ventricular septal defects. Although in such patients both the right bundle branch and the anterior fibers of the left bundle branch were severed, nevertheless it may be expected that normal A-V conduction will continue in this patient population for many years through uninterrupted left bundle branch fibers oriented toward the posterior part of the left ventricle.

In addition to bifascicular injury causing an electrocardiographic right bundle branch block and left anterior hemiblock pattern, experimental data as well as clinical-pathological data indicate that discrete lesions to the distal (branching) part of the His bundle may also account for an identical electrocardiographic pattern. Although small in size, these lesions are located in a strategic crossroad for A-V conduction adjacent to the remaining intact His bundle fibers. It can then be anticipated that small additional injury due to fibrosis which may develop in later years may cause severe A-V conduction impairment.

Figure 1 depicts the cumulative data reported in eight different series of long-term prognosis for patients operated mostly for correction of tetralogy of Fallot and some for isolated ventricular septal defect. The thrust of these previously reported studies was to review prognosis of patients with primarily postoperative right bundle branch block and left anterior hemiblock patterns and various other types of conduction defects other than isolated right bundle branch block. Although some bias is inherent in any retrospective study, such a review allows for a more accurate impression of long-term prognosis than any of the isolated studies alone.

A total of 1856 patients followed from one month to 14 years are reported. The cumulative incidence of right bundle branch block and left anterior hemiblock pattern for the entire series was 11%; over 4% in these series were noted to have postoperative transient complete heart block and 3% developed transient complete heart block and right bundle branch block and left anterior hemiblock in the immediate postoperative period. Of the 204 patients who developed right bundle branch block and left anterior hemiblock, one developed late complete heart block and two died suddenly for total late complications of 1.5%. Four of the 81 patients who developed transient complete heart block in the immediate postoperative period developed late complete heart block and 16 patients (29%) who had transient complete heart block and right bundle branch block and left anterior hemiblock either developed late complete heart block or died suddenly.

These data strongly suggest that transient complete heart block is a predictor for development of late complete heart block or sudden death at least as strong as right bundle branch block and left anterior hemiblock. Further, transient complete heart block in association with right bundle branch block and left anterior hemiblock is a more powerful predictor for the possibility of either developing late complete heart block or dying suddenly than either bifascicular block or transient complete heart block alone. The data also suggest that transient complete heart block may not be a benign postoperative complication as has been previously suggested. The association of a postoperative bifascicular block pattern and transient complete heart block strongly suggest that in this patient population, the site of injury is most likely to be the distal part of the His bundle as has been previously produced experimentally by a technique similar to that used during open heart repair. The likelihood for
the presence of a distal His bundle lesion in this patient population rather than a true right bundle branch block and left anterior hemiblock is also supported by the observation of Godman et al. that most patients who develop late complete heart block had a "bifascicular block" pattern and in addition prolonged H-V intervals indicating the presence of severe His-Purkinje conduction disturbance.

The group of patients with a distal His bundle lesion and the group of patients with a true right bundle branch block and injury to the anterior left bundle branch fibers who display an identical electrocardiogram of the so-called bifascicular block pattern cannot be differentiated currently by using the scalar electrocardiogram. However, the clinical likelihood for the presence of a His bundle lesion is strongly suggested in postoperative patients who in addition to a bifascicular block pattern display prolongation of the P-R interval or a transient complete heart block in the immediate postoperative period. In the absence of a prolonged P-R interval or transient complete heart block in the immediate postoperative period, detection of a prolonged H-V interval is useful in establishing the state of atriovenricular conduction and the type of injury causing the bifascicular block pattern on the electrocardiogram.

Right Bundle Branch Block and Left Anterior Hemiblock Pattern and P-R Prolongation ("Trifascicular Block Pattern")

This electrocardiographic pattern has been attributed in the past to complete injury of the right bundle branch, left bundle branch fibers oriented toward the anterior part of the left ventricle and in addition, to incomplete injury of the posterior left bundle branch fibers. Whereas injury to the right bundle branch and the anterior fibers of the left bundle branch may be understood easily because of the proximity of these structures to the operative field, injury to the posterior left bundle branch fibers during surgery can hardly be explained on this basis because these latter fibers are not in close proximity to the surgical field carried out virtually always from within the right ventricle.

However, this electrocardiographic pattern may be easily explained on the basis of injury to the distal part of the His bundle causing an electrocardiographic pattern of right bundle branch block and left anterior hemiblock patterns and in addition extension of the same lesion to a more proximal (penetrating) part of the His bundle causing an additional first degree A-V block. Indeed such extensive lesions involving both the penetrating and the branching portions of the His bundle have been produced experimentally and caused a trifascicular block pattern on the electrocardiogram.25

The incidence of trifascicular block pattern in patients following correction of intracardiac defects is smaller than isolated bifascicular block pattern. However, by virtue of the extent of the incurred damage and effect on atrioventricular conduction, patients with this electrocardiographic pattern may be expected to have a higher late incidence of either complete heart block or sudden death in the follow-up period. The limited available data indicate that three of 10 patients with this electrocardiographic pattern died suddenly in the follow-up period, an incidence of 30% similar to patients with transient complete heart block and bifascicular block.

Conclusion

It seems clear that in addition to hemodynamic status, presence of residual defects and state of the myocardium, postoperative prognosis for patients with congenital heart disease is related in part to the presence of cardiac electrical instability. Improvement in surgical techniques brought about a very gratifying state of events in which decreased surgical mortality and significant hemodynamic improvement was obtained almost universally. This state of decrease in nonelectrical complications focused attention on the relative significance of surgically induced injury to the various parts of the cardiac specialized conduction system and its subsequent effect on prognosis.2 9 31-36

It seems clear that increased efforts are required to preserve not only the His bundle but the entire cardiac conduction system during open heart surgery. Additional prospective studies of different patient populations with different types of conduction defects are needed to establish the precise effect of these conduction defects on prognosis.

References

The Anatomical Substrates of Wolff-Parkinson-White Syndrome

A Clinicopathologic Correlation in Seven Patients

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SUMMARY Clinicopathological correlations were made on the hearts from seven patients known to have exhibited electrocardiographic evidence of the Wolff-Parkinson-White syndrome. In each case, clinical and pathological investigations were conducted independently, neither group of investigators having knowledge of the other's results. In all seven hearts, the entire atrioventricular junctions were serially sectioned. Accessory atrioventricular connections were predicted in all seven cases following electrocardiographic investigation. Connections were identified histopathologically in four hearts in the predicted site. In another case two connections were identified, one being considered responsible for the pre-excitation. In the sixth case a right lateral connection was anticipated, but only accessory nodo-ventricular fibers were identified following histopathologic studies. In the final case, a posterior septal connection was predicted but the entire septum had fibrosed following previous operation. These findings are discussed in the light of the investigative techniques used, the theories of pre-excitation and the embryogenetic mechanisms producing accessory atrioventricular connections.

Many clinical and electrophysiological studies of the Wolff-Parkinson-White (WPW) syndrome exist; in comparison, relatively little is known of the underlying pathology. Indeed at present there is still a dispute regarding the precise anatomy that underlies the abnormal ventricular activation pattern found in this syndrome. Paladino¹ and Kent² had described myocardial fibers in normal hearts that connected atrial and ventricular myocardium which they considered to be responsible for normal atrioventricular conduction. However, these findings could not be considered to represent normality since Tawara³ demonstrated the existence of the atrioventricular node and its connection with the atrioventricular bundle,⁴ and then Hering⁵ demonstrated experimentally that in the dog heart this bundle was the only conducting connection between atria and ventricles. In their classic study on the WPW syndrome Holzmann and Scherf⁶ suggested two possible mechanisms, one of which assumed the existence of conducting muscular bridges between atrial and ventricular myocardia. Considerable time elapsed before such accessory pathways were actually demonstrated in a patient with WPW by Wood, Wolfirth and Geckeker.⁷ Since then the number of cases in which these accessory connections have

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