Corrected Transposition of the Great Vessels, Atrioventricular Heart Block, and Ventricular Septal Defect

A Clinical Triad

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Three cases with the triad of ventricular septal defect, atrioventricular heart block, and corrected transposition of the great vessels are presented. In each instance the patient successfully underwent open heart surgery on the pump-oxygenator for closure of the ventricular septal defect. A review of the literature suggests a frequent association of these 3 congenital anomalies. In our experience atrioventricular heart block is rarely encountered with simple ventricular septal defect. The finding of atrioventricular heart block in a patient with an interventricular septal defect warrants consideration of an associated corrected transposition of the great vessels.

CORRECTED transposition of the great vessels is a rare anomaly in which the root of the aorta arises anterior to the pulmonary artery, yet each major vessel receives blood from its “correct” ventricle, (the aorta from the left ventricle, the pulmonary artery from the right). As illustrated in figure 1, the main pulmonary artery fails to arch in front of the aorta in the normal manner, and the left pulmonary artery passes behind the ascending aorta. Figure 2 illustrates the medial displacement of the cardiac catheter as it passes through the main pulmonary artery in corrected transposition. In complete transposition of the great vessels the position of the major trunks is similarly altered but they originate from reversed ventricles, the aorta from the right, and the pulmonary artery from the left ventricle.

We are reporting 3 cases that represent combinations of defects that have been reported previously but whose association has not been stressed, namely “corrected transposition” of the aorta and pulmonary artery, ventricular septal defect, and congenital atrioventricular heart block. The diagnosis was confirmed in all patients at open heart surgery on the pump-oxygenator. All demonstrated a gratifying clinical response after closure of their ventricular septal defects.

Case Reports

Case 1. A 10-month-old white boy was the product of a gestation that was unremarkable except for fetal bradycardia (70 to 80/minute) detected during the seventh month of pregnancy. At 11 weeks of age the infant was admitted to Brooke Army Hospital following treatment for pneumonia and congestive failure at another hospital. Examination revealed a small, feeble baby with a pulse rate of 64 and a respiratory rate of 60, but without cyanosis. There was severe pectus excavatum. Oxygen saturation on an ear oximeter reading was 97 per cent. Cardiac fluoroscopy revealed increased pulmonary vascularity and combined ventricular enlargement. An electrocardiogram revealed complete atrioventricular block (fig. 3).

The infant did poorly, with recurrent congestive failure and repeated pulmonary infections. Cardiac catheterization on October 23, 1956, showed a right-to-left shunt at the ventricular level. The pressure in the right ventricle was 65/0 mm. Hg and 65/20 in the pulmonary artery. Systemic arterial blood was 95 per cent saturated. On November 27, 1956, there was clinical and radiographic evidence of atelectasis of the left lower lobe.

Open heart surgery on the pump-oxygenator

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was performed at the Texas Children's Hospital on December 20, 1956. Exploration revealed a corrected transposition and a ventricular septal defect 2 cm. in diameter located high in the septum behind the right atrioventricular valve. This was closed by figure-of-8 sutures. The patient tolerated the procedure well and had an uneventful postoperative course. At the time of discharge 10 days after surgery, the murmur was inaudible. Continued improvement was noted on follow-up examination 2 months later.

Case 2. This 13-year-old white girl was the product of a gestation complicated by repeated episodes of threatened abortion during the first trimester. Cyanosis was observed in the immediate postnatal period and a murmur was detected at 3 months of age. In the first 3 years, there were recurring episodes of respiratory infection, including 5 bouts of pneumonia in 1 year. During this period the patient manifested continuous cyanosis and had frequent "attacks" characterized by syncope and more severe cyanosis.

At age 8 cardiac catheterization was thought to demonstrate a single ventricle with transposition of the great vessels. Arterial oxygen saturation was 86 per cent. During the following 4 years her general condition improved. She was no longer cyanotic and had increased exercise tolerance.

Physical examination revealed a child of slender build, weighing 56\(\frac{1}{2}\) pounds. There was questionable cyanosis of the fingers and toes, with slight clubbing. The ventricular rate was 90 per minute. The electrocardiogram showed complete heart block (fig. 4). Fluoroscopy showed marked cardiomegaly with a globular configuration of the heart and increased pulmonary vascularity without hilar dance. The pulmonary artery segment was not prominent (fig. 5). An oximeter reading was 95 per cent.

Cardiac catheterization revealed a large ventricular septal defect with predominantly left-to-right shunt. There was an increase from 61 per cent to 80 per cent saturation of blood from the right atrium to the right ventricle; femoral arterial blood saturation was 95 per cent; right ventricular pressure was 115/10 mm. Hg.

At operation there was corrected transposition of the great vessels and a 3-cm. defect high in the membranous septum. The defect was repaired. The patient tolerated the procedure well and there was a demonstrable reduction in pulmonary artery pressure after closure of the defect. The patient has continued to do well after discharge from the hospital.

Comment. The progressive decrease in cyanosis and improvement in exercise tolerance during several years prior to surgery will be the subject of a later report following postoperative catheterization, since it raises the question of possible cardiac surgery at a later date in cyanotic children with septal defects. In the past cyanosis has been considered to represent an irreversible process of increased pulmonary vascular change. This child had well-documented persistent cyanosis until 8 years of age. Then her cyanosis virtually disappeared and at 12 years of age, pulmonary vascular resistance had diminished to the point that pulmonary blood flow exceeded systemic flow. The child appeared to be benefited by closure of her ventricular septal defect.

Case 3. This 6-year-old white boy had a known cardiac murmur at 5 weeks of age. He was dyspneic on exertion. Physical examination revealed him to be well developed and nourished and without cyanosis. There was slight bulging of the left anterior chest wall. The electrocardiogram revealed incomplete atrioventricular block with intermittent dropped beats (fig. 6). Roentgenograms and fluoroscopy revealed an enlarged globu-
CORRECTED TRANSPOSITION

Fig. 2. Position of the cardiac catheter in the normal heart (Left) compared with that in corrected transposition (Right). In each case the catheter tip lies in the right pulmonary artery. The more medial position of the pulmonary outflow tract in corrected transposition can be seen by the relatively greater distance from the catheter to the left border of the cardiac silhouette.

lar heart with increased pulmonary vascular markings. Cardiac catheterization showed a left-to-right shunt at the ventricular level and pulmonary hypertension (90/50 mm. Hg). On November 27, 1956, the ventricular defect was closed. The patient was noted to have corrected transposition of the great vessels. During the early postoperative period transient atrioventricular dissociation was observed. On the third postoperative day the patient developed abdominal distention and liver enlargement, but there was prompt improvement following the administration of digitalis and mercurial diuretics. The patient was discharged from the hospital in good condition on December 16, 1956.

DISCUSSION

Corrected transposition is a rare anomaly. Abbott in 1936 reported a total of 6 cases in her series of 1,000 congenital defects. Considerable difference of opinion has been expressed by anatomists and embryologists concerning the nature and evolution of this defect. This is rather extensively reviewed by Bremer,2 Harris and Farber,3 Lev and Saphir,4 and Cardell.5 It must be understood that the term corrected transposition refers to a functional “correction.” Despite the varieties of transposition and inversion that the great vessels, atria, and ventricles may undergo, the end result is that the aorta receives oxygenated blood and the pulmonary artery receives systemic venous blood. Cardell5 cites 8 different anatomic combinations that may occur in corrected transposition.

In the usual form of corrected transposition the aorta arises anterior and to the left of the pulmonary artery. The left ventricle is continued upward and anteriorly into a short infundibular portion from which the aorta arises. There is no infundibular portion of the right ventricle and the pulmonary artery arises directly from the base of this ventricle in close proximity to the usual location of membranous ventricular septal defects. As pointed out by Edwards,6 “The architecture of the right ventricle resembles that of a normal left ventricle and, conversely, that of the left-sided ventricle resembles the architecture of a normal right ventricle. The right atrioventricular valve in corrected transposition resembles a normal mitral valve in its structure, whereas the left atrioventricular valve structurally resembles a normal tricuspid valve.” The left-sided tricuspid valve is frequently deformed and incompetent in corrected transposition. The aorta and pulmonary artery arise side by side without the normal spiraling and the left pulmonary artery passes posterior to the ascending aorta. Functionally corrected transposition is compatible with normal longevity. It is the frequently associated anomalies that cause the morbidity and mortality in this disease.

In 1956 Cardell5 reviewed the literature on corrected transposition and reported a case with corrected transposition, ventricular septal defect, and 2:1 atrioventricular block. In a review of 18 cases of corrected transposition from the literature he reported an
equal sex distribution. Duration of life varied from 9 days to 46 years (average 14 years). Septal defects (atrial or ventricular or both) were especially common, and in only 4 cases were both septa intact. Only 2 cases had no associated anatomic defects, and 1 of these had complete heart block. Of the 18 cases from the literature with adequate records, 3 had complete heart block and a fourth 2:1 A-V block. Helmholtz et al., whose paper was not included in Cardell’s review, reported 1 patient with complete heart block who demonstrated corrected transposition at catheterization. In addition these authors reported 6 cases of corrected transposition studied pathologically. Three of these had associated ventricular septal defects, 5 had defects of the left atrioventricular valve, 3 had anomalous insertion of the chordae, and 2 had a left-sided Ebstein type of malformation. None of the 6 cases was reported to have atrioventricular block but this was a pathologic rather than a clinical study and this information may not have been available. Donoso et al. also reported a case not included in Cardell’s review (case 1 in their report), who had congenital complete heart block, corrected transposition, ventricular septal defect, and atresia of the mitral valve.

In 1933 Yater et al. gathered 44 examples of congenital atrioventricular heart block from the literature and reported 1 case of congenital complete atrioventricular block with ventricular septal defect studied by serial sections through the conduction system. Of the 44 cases with atrioventricular block, 29 had associated ventricular septal defects. Corrected transposition was not mentioned in this paper. However, a review of figure 4 shows that in the case they reported the aorta arose anteriorly, and we believe it also represents an example of the triad. Yater and Edwards have reviewed the published photographs and agree that this is probably true.

Keith et al. reviewed 44 necropsied cases of complete transposition of the great vessels. They found patent foramen ovale in all, patent ductus arteriosus in 25, ventricular septal defect in 18, and atrial septal defects in 2. Electrocardiograms were taken in 30 cases but no comment was made on the presence of conduction defects. Kjellberg et al. presented 8 cases of complete transposition, 3 with associated ventricular septal defects, 3 with patent foramen ovale, and 1 with atrial septal defect. Electrocardiograms were done in 7 instances, but conduction defects were not remarked upon. This suggests that complete transposition of the great vessels is less
commonly associated with atrioventricular block than is corrected transposition. It is obvious that in complete transposition, survival beyond the immediate postnatal period necessitates the presence of a shunt to permit some of the oxygenated blood to perfuse the systemic vessels. This is not essential for survival with corrected transposition. However, it appears that ventricular septal defects are equally common in both conditions.

Although the association of congenital heart block with ventricular septal defect has been stressed in the past, it should be noted that in the 200 cases of interventricular septal defect reviewed by Brown, no cases of congenital heart block were found. Downing and Goldberg reviewed 100 cases of ventricular septal defects. Electrocardiograms were done in 91. There were no instances of complete heart block, although incomplete atrioventricular block was found in 3. Kjellberg et al. cite 54 cases of ventricular septal defects with no associated complete heart block although 4 presented delayed atrioventricular conduction. In neither series was corrected transposition reported.

Of 69 patients operated at the Texas Children’s Hospital for closure of ventricular septal defect, 3 had associated corrected transposition of the great vessels. All of these had associated congenital atrioventricular block; this was complete in 2 instances and incomplete in 1. One additional patient had transposition of the great vessels with a common ventricle; this patient did not manifest atrioventricular block. Of the remaining 65 patients with ventricular septal defect and normal origin of the great vessels none had complete atrioventricular block, only 4 manifested incomplete atrioventricular block, and in only 1 of these did the P-R interval exceed 0.20 second. Lillehei has also observed the association of ventricular septal defect,
corrected transposition of the great vessels, and complete atrioventricular block. Whereas acquired atrioventricular block may be a serious complication of open cardiac surgery for repair of ventricular septal defects, experience indicates that congenital atrioventricular block is usually well tolerated.

SUMMARY
Three cases with the triad of atrioventricular heart block associated with ventricular septal defect and corrected transposition of the great vessels are reported. All cases underwent open heart surgery on the pump-oxygenator for closure of their ventricular septal defect. A review of the literature suggests that atrioventricular heart block is associated more frequently with corrected transposition of the great vessels and ventricular septal defect than with isolated ventricular septal defect or with other congenital anomalies. The finding of congenital atrioventricular block in association with ventricular septal defect should alert the observer to search for corrected transposition of the great vessels.

ADDENDUM
Dr. Howard B. Burchell of the Mayo Clinic has also observed 3 cases of this syndrome and believes that there is evidence to support the viewpoint that there is sufficient consistency in the association of the finding of heart block, corrected transposition, and ventricular septal defect to constitute a clinical syndrome.

SUMMARIO IN INTERLINGUA
Es reportate 3 casos del triada de blocco cardiaco atrioventricular con defecto ventriculoseptal e corrigite transposition del grande vasos. Omne li cases esses subiekte a chirurgia cardiaco apertae, con le oxygenator-pumpa, pro effectuare clausione del defecto ventriculoseptal. Un revista del litteraturae pare indicar que blocco cardiaco atrioventricular est associate plus frequentemente con transposition corrigite del grande vasos e defecto ventriculoseptal que con defecto ventriculoseptal sol o que con altrae anomalias congenite. Le constatazione de congenite blocco atrioventricular in association con defecto ventriculoseptal debeerea alamar l'observer a cercar transposition corrigite del grande vasos.

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