CASE REPORT

Hypoplastic and Fibrotic Sinus Node Associated with Intractable Tachycardia in a Neonate

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SUMMARY A case of fetal and neonatal persistent atrial tachycardia is described in a child with complete transposition of the great arteries. At autopsy, serial sections of the conducting tissue showed that the sinus node was hypoplastic and markedly fibrotic. The sinus node consisted of a small area of cells clustered around the nodal artery. Although the underlying mechanism of neonatal persistent tachycardia is unclear, it is possible that the hypoplasia and fibrosis of the sinus node was the substrate in the present case. This mechanism should be considered in any newborn infant in whom sinus rhythm cannot be established.

CHRONIC supraventricular tachycardia is occasionally seen in infants and children both with and without structural heart disease.1,2 Presentation may be with symptoms and signs associated with an atrial tachycardia alternating with short periods of sinus rhythm3 or with a chronic persistent atrial tachycardia that seldom, if ever, gives rise to sinus rhythm.4 Recently, intracardiac electrophysiologic studies have defined mechanisms of supraventricular tachycardia that include both enhanced automaticity and reentry circuits.5,6 Though the electrophysiologic mechanisms are often well known, there have been few clinicopathologic studies, as these arrhythmias are rarely fatal. We recently studied a fatal case of chronic atrial tachycardia in a newborn infant with fetal tachycardia and complete transposition of the great arteries. In this report, we present some unusual abnormalities of the conduction system.

Case Report

The mother was induced at 39 weeks gestation by artificial rupture of membranes (infant birth weight 2.8 kg). Before induction on admission to hospital the baby had a persistent tachycardia and an emergency lower-section cesarean section was carried out. He was floppy and deeply cyanosed at birth and spontaneous respiration commenced only after 7 minutes of intermittent positive pressure respiration via an endotracheal tube. The heart rate was 180 beats/min. The infant remained tachypneic and cyanotic and was transferred to Hammersmith Hospital at 2 days of age. He was not in congestive cardiac failure. The cardiac impulse was right ventricular in character. The second heart sound was loud and single and there were no murmurs. All the arterial pulses were palpable and the systolic blood pressure in the upper and lower limbs was 60 mm Hg. He had a regular tachycardia of 190 beats/min and his ECG showed atrial flutter with 2:1 atrioventricular block (fig. 1). In addition there was right-axis deviation and right ventricular hypertrophy. Chest x-ray showed his heart size to be at the upper limit of normal, with normally filled lung fields. His pH was 7.06, Pco2 47 mm Hg and Po2 25 mm Hg. A diagnosis of transposition of the great arteries was made and transfer to the cardiac catheter laboratory arranged. The diagnosis was confirmed and a ventricular septal defect was excluded by angiocardio- graphy. The ductus arteriosus was patent. An atrial septostomy was performed and the Po2 rose to 40 mm Hg. Thereafter the infant's color remained satisfactory and he was less tachypneic, but his heart rate continued to be about 200 beats/min. Over the next 14 days he continued to have a persistent atrial tachycardia despite successive treatment with verapamil, digoxin, propranolol, disopyramide and DC cardioversion. Combinations of digoxin with propranolol and disopyramide were also unsuccessful in obtaining sinus rhythm. Both atrial flutter with varying degrees of atrioventricular block and chaotic atrial tachycardia occurred after balloon atrial septostomy (fig. 2).

At the age of 1 week he had a respiratory arrest associated with aspiration of a nasogastric feed and from then until his death he failed to breath spontaneously.

The heart and lungs were examined at autopsy. The segmental arrangement was situs solitus, atrioven- tricular concordance and ventriculoarterial discordance (complete transposition). The aortic valve was right-sided relative to the pulmonary valve and there was a patent ductus arteriosus, atrial septal defect and double orifice of the tricuspid valve. The entire atria and adjacent parts of ventricular myocardium were serially sectioned (fig. 3); initially, one section in twenty-five cuts was mounted and intermediate sections in areas of special interest were mounted subse-
The atrial tissues were hypertrophied and showed considerably more fibrous matrix than normally found at this age, especially at the crista terminalis. There was also considerable thickening of the subendocardial layers, with a dense fibroelastic layer. The sinus node was poorly formed and was well posterior, being represented by only a small area of cells clustered around a prominent artery (fig. 4). The area was considerably smaller than the normal node (compare figures 4A–4C). The quantity of fibrous tissue within the hypoplastic nodal area was also considerably greater than normally encountered (fig. 5). The margins of the hypoplastic node merged with the

“working” atrial myocardium, which was fibrotic but otherwise not unusual. The atrioventricular node was a well-formed structure adjacent to the annulus fibrosus, and was normally situated relative to a well-formed tendon of Todaro. It was centrally placed within the septum, with both right and left extensions of the compact node present. There was a well-formed nodoventricular fiber between the compact node and the crest of the ventricular septum (fig. 5A). The penetrating bundle entered the middle segment of the muscular septum, with no atrioventricular segment of the membranous septum visualized. The bundle penetrated some distance intramyocardially before veering to the left (fig. 5B). The bundle ran an extensive intramyocardial course before emerging subendocardially and ramifying as the left bundle branch, which was poorly formed. The right bundle branch was searched for but not identified.

Discussion

Although considerable advances are being made in the understanding of the underlying mechanisms of supraventricular tachycardia, knowledge of the pathologic basis is often lacking. Unfortunately, the majority of children who reach autopsy usually do not have a detailed examination of their conducting tissue. Brechenmacher and colleagues described the histologic features found in an infant with intractable tachycardia. At autopsy, abnormalities in the atrioventricular conduction system were present but, as in the present case, the authors were only able to

**Figure 1.** ECG (lead II) showing atrial flutter with 2:1 atrioventricular block.

**Figure 2.** ECG showing chaotic atrial tachycardia with right bundle branch block. Both the p-wave morphology and axis are continually changing. Right bundle branch block developed after the balloon atrial septostomy unrelated to specific drug therapy. The fourth beat of the simultaneous three-channel recording probably arises in or around the sinus node. This does not necessarily suggest that there was sinus automaticity, but is consistent with isolated reentry from either manifest or concealed atrial extrasystole. The latter is more consistent with the histologic findings. Review of several thousand complexes recorded throughout this infant’s life did not reveal any other similar complexes.

**Figure 3.** A drawing of the right atrium and ventricle of the heart, showing the block removed from serial sectioning, the plane of section and the position of the sections illustrated in figures 4 and 5.
speculate as to the relationship between the abnormalities found in the conducting system at autopsy and the cardiac arrhythmias present during life. Nevertheless, studies such as these demonstrate the additional information available from each patient in whom the conducting system is carefully examined and provide clues to the underlying structural abnormalities.

Figure 4. Sections through the hypoplastic sinus node (panels A and B) compared with a normal infant sinus node in a similar plane of section (panel C). Note the severe hypoplasia of the nodal tissue of the present case. Trichrome stain, low-magnification views.

Figure 5. Photomicrographs comparing the morphology of the normal node (panel A) with the hypoplastic node in the present case (panel B), showing the greater proportion of fibrous tissue in the hypoplastic node. Trichrome stain, high-magnification views.
It would, however, be very unlikely that sinus rhythm could ever occur where the sinus node is represented by a small area of cells, particularly when the area also shows marked fibrosis. Hypoplasia and fibrosis of the sinus node have not, to our knowledge, been previously described, though this may be due more to the relatively small number of detailed pathologic studies than to the absolute rarity of the disease. It would be an appealing hypothesis to suggest failure of normal development of the sinus pacemaker as a cause of chronic atrial tachycardia in neonates and infants, but the fibrosis in both the hypoplastic node and atrial myocardium must not be ignored as an alternative mechanism.

During the development of the conducting system of the heart there is a sequence of pacemaking tissues. The first detected is in the primitive ventricle, followed by the atrial myocardium. Later, the atrial myocardium is supplanted by the fast intrinsic rate of the cells that originate from the sinus venosus and become the sinus node. Failure of these cells to form a normal specialized pacemaking unit could result in the atrial myocardium persisting as the dominant pacemaking unit. Chronic atrial rhythm may then be expected to be present from birth. The excessive amounts of fibrous tissue in the atrial myocardium would have exacerbated not only the problem of pacemaking but also of atrial conduction. In our patient, sinus rhythm was not recorded at any time and all ECGs showed that cardiac conduction started in the atria. Normal sinus rhythm would not be expected at any stage during fetal development and in our patient, tachycardia was noted clinically on admission to hospital before induction and was recorded at the time of artificial rupture of membranes when the fetal ECG was first recorded.

Patients with supraventricular tachycardia may have abnormal sinus function. If verapamil or propranolol had been successful in converting our patient's tachycardia to sinus rhythm, the added effects of these drugs on an already hypoplastic sinus node might have led to complete cardiac arrest.

In conclusion, a case of chronic atrial tachycardia is described in which the underlying mechanism was probably due to severe hypoplasia and fibrosis of the sinus pacemaker. Control of the conduction system was by the more primitive atrial myocardium, which was itself fibrotic. The role of this mechanism in the development of neonatal persistent tachycardia is unclear but warrants further investigation.

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References

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