Spinnaker Formation of Sinus Venosus Valve

Case Report of a Fatal Anomaly in a Ten-Year-Old Boy

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SUMMARY
A persistent right valve of the sinus venosus fatally complicated a case of tricuspid atresia in a 10-year-old boy. The center of this large membrane stretched to form a sac resembling a spinnaker. Depending on the relative amounts and directions of blood flowing against it from the coronary sinus below and the venae cavae above, the sail might fill or empty quickly. This gave means of potential obstruction: either of coronary venous flow which, without a tricuspid valvular orifice, could be trapped beneath the membrane; or of all right atrial flow if the sac fell across the sole atrial outlet, the septal defect. Such episodic block caused syncope six times in the last 23 months of life and death in acute right heart failure.

Additional Indexing Words:
Tricuspid atresia  Obstruction, episodic  Intermittent syncope

POSTNATAL LIFE with tricuspid atresia requires patency of an alternate route by which systemic and coronary venous blood may reach the lungs. There are always an atrial septal defect and either an interventricular or a supraventricular communication. Any further impairment of this abnormal passage is a serious complication and all surgical treatment aims at increasing pulmonary blood flow. Because of its length and deformity the route from the left atrium to the pulmonary artery is particularly liable to obstruction. Therefore when a patient with tricuspid atresia gives evidence of acute circulatory failure, one should consider first in diagnosis, interruption of that tenuous connection.

Report of Case
K. S., born September 24, 1955, was an apparently normal male infant, the product of an uncomplicated pregnancy and delivery. Increasing cyanosis, first noted at 2 months of age and later accompanied by dyspnea, prompted his admission to the University of Oregon Medical School Hospital at 2½ months. The liver was enlarged and pulsatile, and the spleen was readily palpable. A diagnosis of tricuspid atresia or stenosis with interatrial, and possibly interventricular, septal defects was made and a Potts aortico-pulmonary anastomosis was performed at the age of 4½ months. The cyanosis then decreased markedly, and evidence of failure disappeared. However, his subsequent growth rate was subnormal; his height and weight were below the third percentile at the age of 8 years, 11 months. At that time he came to Congenital Heart Clinic having had three occurrences of syncope in the past 6 weeks. Two of these had occurred during exertion, the third at night. All were momentary and without sequelae.

He was then hospitalized, cyanotic, with clubbing of all digits, and in congestive failure. The liver edge was 4 cm below the right costal margin and the hematocrit was 66%. The physical and laboratory findings, supported by cardiac catheterization (table 1) and angiocardiography, signified that flow through the anastomosis was inadequate. A small interventricular septal defect was seen leading into the rudimentary right ventricle; there was no ductus. There were elevated right atrial pressure and low oxygen saturation in the coronary sinus. A Glenn procedure (diversion of the superior vena cava into the right pulmonary artery with closure of the caval ostium in the right atrium) was performed. The postoperative course was uneventful. Cyanosis and clubbing decreased; arterial oxygen saturation rose to 87%.

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Table 1
Cardiac Catheterization after Potts Anastomosis

<table>
<thead>
<tr>
<th>Oxygen saturation % (by cuvette)</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>63.2</td>
</tr>
<tr>
<td>Right atrium</td>
<td>65</td>
</tr>
<tr>
<td>Coronary sinus</td>
<td>19</td>
</tr>
<tr>
<td>Left atrium</td>
<td>81.2</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>84.6</td>
</tr>
<tr>
<td>Subclavian artery</td>
<td>83.2</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>80</td>
</tr>
</tbody>
</table>

At the age of 10 years, 8 months, the patient had two more brief fainting episodes 6 days apart and was admitted again. He related having had tingling of the feet, headaches, and abdominal pain in association with the last event. Blood pressure was regularly 94/50 mm Hg, pulse 96, and respirations 20. Exercise tolerance was reduced. Investigation disclosed no additional abnormalities, and the syncope was regarded as due to either hypotension or hypoxia, intermittent and with etiology undetermined. He was discharged after 9 days.

Twenty-eight days later, while playing base-

ball, he was suddenly forced by weakness to lie down. He vomited and was brought to the hospital emergency room. He was awake, cyanotic, in acute distress, and occasionally gagging. Blood pressure was 40-50/0 mm Hg, pulse 160 per minute, respirations 30, and temperature 102.2 F. The liver extended 2 fingerbreadths below the costal margin and was tender. Cranial nerve functions were normal and Babinski reflexes absent; there was normal response to pain. Therapy included an oxygen tent, digitalis, steroids, and intravenous Levophed, Keflin and penicillin (20 million units). Tracheostomy, positive-pressure respiration, and peritoneal dialysis were applied, the last when serum levels of potassium reached 7.2 mEq/L. Arterial oxygen saturation was 93% and venous 30%; blood pH was 7.36. White blood count, 11,000/mm³ on admission, reached 47,000 terminally. The potassium level rose to 8 mEq/L and blood pH fell to 7.16. Heart rate varied from 40 to 120 with atrial fibrillation and nonspecific ST abnormalities. The temperature dropped to 97.2 F, and another dialysis returned little potassium owing to poor perfusion. The heart stopped permanently 50 hours after admission.

Figure 1

Left. Open right atrium, posterior view. Persistent right sinus venosus valve, based mostly on the crista terminals, is collapsed against the trabeculated portion of the atrium and out of view (arrow). The coronary sinus (interrupted oval) and atretic tricuspid valve are below this membrane and also out of view. A polyethylene tube and rubber balloon have been placed beneath the sinus venosus valve and are protruding from the cut edge of the great coronary vein. Right. The balloon has been filled via the tube expanding the spinnaker-like valve. The mass fills the atrium. Note small segment of balloon (arrow); this signifies the only route of escape of blood from the valve pocket. IVC, inferior vena cava; L SVC, ligated superior vena cava; ASD, atrial septal defect.

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Autopsy Findings and Correlation

The heart weighed 280 g (normal 116 g), the largest proportion being the left ventricle. A smooth fibrous atrial floor replaced the tricuspid valve orifice. The closed superior vena cava ostium and the anterosuperior portion of the right atrial wall in the area of the Glenn procedure were slightly contracted by scar tissue, which included the cephalic part of the crista terminalis. Within the atrium lay a large, smooth-surfaced flap of fibrous tissue with a thin, redundant, distensible center and a tough, cord-like free margin; this flap almost completely subdivided the atrium. Its base measured 6.3 cm, the left portion being in the normal positon of the eustachian valve and separating the ostium of the inferior vena cava from the dilated coronary sinus. The major (superior) portion of the base was sited on the crista terminalis. The length of the free margin was 2.8 cm. Thus blood from the coronary sinus had been forced to take an angulated course from below this membrane to the atrial septal defect. Even without distention this septum across the right atrium was 85% complete. Anterior to it lay the trabeculated portion of the atrium, the coronary sinus, and the site of the atretic tricuspid valve.

The septal defect (secludum type) and the ostium of the inferior vena cava were posterior and superior. The center of the flap when distended was 2.5 cm from the coronary sinus. This sail, 3.8 cm in diameter when filled with blood, almost entirely occupied the nontrabeculated portion. In this state it would have impeded atrial filling, and was in position to cause partial or complete obstruction of the only right atrial outlet, the septal defect, which had a diameter of 1.5 cm. Falling slightly more anteriorly it could have closed off the anterior portion of the atrium and obstructed coronary venous return without affecting caval flow (figs. 1, 2, and 3, lower right).

The pulmonic valve, and especially the right ventricle, were hypoplastic to a degree consistent with the tricuspid atresia. The pulmonary artery, also small proximally, led into larger branches, of which the right had been surgically closed. Microscopically all intrapulmonary vessels were normal. The great arteries were neither transposed nor inverted. There was a high interventricular septal defect 0.2 cm in diameter. The ductus arteriosus was closed, the Potts anastomosis 0.5 cm in diameter and the Glenn 1.5 cm. The liver and spleen had severe acute and chronic passive congestion. Proximal renal tubules showed acute degeneration typical of that ascribed to the use of Levophed in shock. Conspicuous basophilic degeneration involved occasional isolated myocardial fibers.

In postmortem review the several categories of syncope were pondered. Besides cardiac causes, Adams and Harrison classify peripheral circulatory (including psychogenic), chemical, cerebral, and emotional,2 none of these latter fits the evidence. With primary disease of the myocardium and of the cardiac conduction system ruled out, only acute obstruction of cardiac blood flow accounts satisfactorily for the entire clinical and pathological picture. The anomalous sinus venous valve was the only means of accomplishing this intermittently. Presumably it partially obstructed at least coronary venous circulation 2 years before death. The low oxygen saturation in the coronary sinus reflected sluggish circulation therein. In view of the physical nature of this flap (fibrous, thin, pliable) corrective surgery would have been simple at any time. Originally spasmodic, this obstruction became protracted in the last 50 hours of life. Parenchymal hepatic, and renal degeneration caused the terminal fever, hyperkalemia, and acidosis. We did not determine the reason for the inadequate flow through the Potts anastomosis, of which the diameter was sufficient.

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Figure 2

Drawings of similar views, slightly more from the left than figure 1, show possible obstruction of all right atrial flow at the septal defect or of coronary flow only by trapping beneath the valve. Arrow indicates circuitous route of coronary venous blood to septal defect. With the sail filled there is little possibility of any passage through the atrium.
Upper left. At 5 weeks' gestation the right horn of the sinus venosus is prominent and opens into the common atrium on the right side between the right and left sinus venosus valves. Upper right. By 8 weeks the right horn is incorporated into the atrial wall (sinus venarum) and the SVC, IVC, and CS are separated into different openings by the superior and inferior limbic (subendocardial fibromuscular) bands. The left sinus venosus valve is being adsorbed into the interatrial septum and the right normally regresses. Lower left. After 12 weeks the only remaining portions of the septum spurium (right valve) are the crista terminalis and the eustachian and thebesian valves. The inferior limbic band has contributed the transverse parts of these valve bases. Lower right. The present case. The thebesian valve portion has largely regressed but the rest of the septum spurium and the inferior limbic band remain. The atrium is bilocular. Its posterior (smooth-walled) portion includes the ostia of the venae cavae (superior ligated), the interatrial septum and the defect. The anterior portion contains the coronary sinus and the site of the atretic tricuspid valve. The anomalous valve was probably not dilated in early childhood but flow patterns which may have contributed to that dilation are illustrated.

Discussion

Patients with tricuspid atresia commonly die in early childhood but reports exist of survival to older ages and we have performed the autopsy of one such patient who died at 26 years. Symptoms and longevity vary according to the volume of pulmonary blood flow and the Glenn operation, by increasing this, has greatly improved prognosis. This patient profited much from the procedure and might have lived to advanced age had it not been for the additional and unsuspected deformity.

Anomalies of the sinus venosus valves and
morphological variations of their derivatives, the septum spurium, the crista terminalis, and the eustachian and thebesian valves, have been recorded by Leo6 and others7-11 beginning in 1886. However, with one exception, no distortion of any of these structures has previously resulted in cardiac malfunction, although Chiari’s network loomed as a potential source of thrombotic emboli.8,9, 12 In 1957 Rossall and Caldwell12 reported a case of inferior vena caval obstruction by a persistent eustachian valve in an adult with no other congenital anomalies. The lesion here presented is unique. It appears to be a product of unusual persistence of the right sinus venosus valve and of the inferior limbic band.14 Excessive leftward traction of the inferior end of that tissue may also have contributed to the structure (fig. 3).

Lack of a tricuspid orifice compelled coronary venous blood to escape from the blind atrial subdivision by a route which changed direction sharply. This channel was narrow where it rounded the inelastic free edge of the flap. Considerable flow was probably directed at the center of the sail, progressively distending and weakening this part of it.

By reducing atrial inflow, and presumably pressure above the flap, the Glenn procedure possibly increased the effect of the coronary stream in expanding this center (fig. 3, lower right). As the membrane stretched into the shape of a spinnaker its potential for hindering atrial circulation grew. It became capable of blocking either of two narrow channels; the distended pouch, being somewhat movable on its base, abutted against either the atrial wall anteriorly or the septal defect posteromedially. In the one position it might critically hinder coronary venous flow while allowing free passage from the inferior vena cava to the septal defect; or, by a slight shift, it might block the atrium at the defect. Until death some circulation continued in both venous systems. Arrest of inferior caval flow would account for most of the findings; stagnation of coronary venous flow might have elicited the severe myocardial degeneration seen microscopically. Intermittent cessation of flow therein might have rendered the myocardium acutely incapable of adequate contraction.

Acknowledgment
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References

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