Stenotic Semilunar Valve in Persistent Truncus Arteriosus*

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The clinical, hemodynamic, and pathologic findings in two newborn infants with persistent truncus arteriosus and stenosis of the truncal valve are described. In one case the anatomic features of the basic condition were classic, with a dysplastic semilunar valve which was mainly stenotic and also incompetent, while in the other the truncus arteriosus arose exclusively from the right ventricle and was almost exclusively stenotic. A ventricular septal defect was the only outlet for the left ventricle. In this case, mitral stenosis was also present and associated with a left-to-right shunt at the atrial level.

The valve of persistent truncus arteriosus is often dysplastic and incompetent but rarely stenotic. The clinical, hemodynamic, and pathologic findings of a stenotic truncal valve were first described in a case report by Burnell and associates in 1971 and again by Lee and associates in 1973. We have observed two examples of truncal valvular stenosis; our case 1 was similar to that of Burnell et al. and case 2 had some findings like that of Lee et al.

The purpose of this report is to present the clinical, hemodynamic, and necropsy findings of these two cases. Both had been admitted shortly after birth (within a three-week period) to the intensive care nursery of St. Mary's Hospital, Madison, Wis. We could not find any common features in the gestational histories to indicate pathogenesis. However, case 1 had a strong family history of congenital heart disease.

Following initial management of congestive failure, both babies were subjected to cardiac catheterization to establish the diagnosis. Thereafter, they were transferred to the University of Minnesota Hospitals and the Mayo Clinic, respectively, for further consideration regarding the feasibility of operation.

CASE REPORTS

CASE 1

Clinical Features. The boy was the product of an uncomplicated 42-week gestation in a 27-year-old primigravida woman. Delivery was uncomplicated, and the infant's birth weight was 4,026 gm (8 lb, 14 oz). Resuscitative measures were not required, and the Apgar score was 9 at one minute. However, at 30 minutes the skin had become dusky; and tachycardia, tachypnea, and a loud continuous murmur were present.

Cardiac consultation at 75 minutes revealed mild cyanosis of the lips while the infant breathed high concentrations of oxygen. The respiratory rate was 70/min, with intercostal retractions. A coarse thrill was palpable at the apex and left axilla. Both heart sounds were loud. A grade 3/6 systolic murmur was followed by a single second sound. A loud high-pitched murmur persisted through diastole, was loudest at the cardiac base and third left interspace, and sounded more like a ductus arteriosus than aortic insufficiency. A loud diastolic flow murmur was heard at the apex. The brachial and femoral pulses were normal. The hepatic edge was palpated 3 cm below the right costal margin.

An electrocardiogram on the day of birth showed sinus rhythm with a rate of 150 beats per minute. The mean frontal-plane QRS axis was +90. The tracing was within the normal range for the newborn. Thoracic roentgenograms showed a shift of the mediastinum and the heart to the left, with prominent pulmonary vascular markings. The cardiac size and the pulmonary congestion increased over the next three days.

Administration of digoxin and ethacryninc acid was begun at 12 hours of age without appreciable response. Cardiac catheterization was performed at 20 hours of age. The catheter could be advanced into the right pulmonary artery from the ascending aorta, suggesting the presence of truncus arteriosus. A withdrawal tracing from the right pulmonary artery to the truncus arteriosus and the right ventricle showed a stepwise increase in pressures (Table 1). The left ventricle...
Table 1—Cardiac Catheterization Data from Cases 1 and 2

<table>
<thead>
<tr>
<th>Site</th>
<th>Case 1 (Age, 20 hr)</th>
<th>Case 2 (Age, 6 days)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Site</td>
<td>Oxygen Saturation (%)**</td>
<td>Pressure (mm Hg)†</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>68</td>
<td>8/3</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>65</td>
<td>6/2</td>
</tr>
<tr>
<td>Right atrium</td>
<td>65</td>
<td>7/1</td>
</tr>
<tr>
<td>Left atrium</td>
<td>93</td>
<td>9/1</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>64</td>
<td>126/9</td>
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<tr>
<td>Left ventricle</td>
<td></td>
<td>(ed = 10)</td>
</tr>
<tr>
<td>Aorta</td>
<td>83</td>
<td>130/5</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>69</td>
<td>72/32</td>
</tr>
</tbody>
</table>

*All venous pressures were "a" waves.
**Saturations were obtained while breathing air.
†Systolic/diastolic; and ed, end-diastolic.
‡Simultaneous pressures.

Figs 1 and 2 (case 1). a (left). Catheter is in dilated truncus arteriosus. Right aortic arch is present. Large pulmonary arteries arise from base of common trunk. b (right). Injection during systole into base of left ventricle. Jet of contrast material streams through stenotic truncal valve.

The family history was interesting in that the infant's father had tetralogy of Fallot with pulmonary atresia, a diagnosis which had been established by cardiac catheterization; also, a paternal aunt had a ventricular septal defect which had been repaired surgically.

Pathologic Features. The cardiac and visceral configuration was that of situs solitus. The heart was moderately enlarged. Only one great vessel, measuring 12 mm in diameter, arose above the ventricles. The pulmonary trunk came off from the left side of this vessel, a situation seen in type 1 persistent truncus arteriosus. The pulmonary trunk divided immediately into the left and right pulmonary arteries with normal course and distribution (Fig 2).

The truncal valve was bicuspid and dysplastic. Both of the thickened cusps showed further incomplete division, suggesting a quadricuspid valve. The diameter of the lumen through the valve was 6 mm (Fig 3). The first set of branches from the truncus arteriosus were the two coronary arteries. They came off behind the opposite, incompletely divided cusps anteriorly and posteriorly. The distribution of the artery arising anteriorly was like that of the right coronary artery, while that of the other was like the left coronary artery. The next branch from the truncus was the pulmonary trunk, 6 mm in diameter, which arose 5 mm above the truncal valve. The aortic arch was right-sided. The branching from the arch had a mirror-image distribution (Fig 2a). The ductus arteriosus was absent. The systemic and pulmonary venous returns were normal, and the coronary sinus opened normally into the right atrium. A 10 X 3 mm atrial septal defect resulting from a valvular incompetent foramen ovale was present at the fossa ovalis. The tricuspid and mitral
Figure 2 (case 1). a (left). Truncus arteriosus has been sectioned above truncal valve to display short pulmonary trunk from which, in turn, right (RP) and left (LP) pulmonary arteries arise. Aorta (Ao) proceeds upward as right aortic arch which lies to right of trachea (Tr). b (right). Right ventricle (RV) and truncal valve. Immediately beneath valve is characteristic ventricular septal defect. (D).

valves were normal and led into hypertrophied and dilated right and left ventricles, respectively. The right ventricular wall measured 5 mm and the left ventricular wall 6 mm in thickness.

The truncus arteriosus was the outlet for both ventricles. It overrode the ventricular septum, arising approximately two-thirds above the right and one-third above the left ventricle (Fig 2). The ventricular septal defect, 10 mm in diameter, was in the superior aspect of the septum and immediately below the origin of the truncus arteriosus. On the right ventricular aspect the poorly developed parietal limb of the crista supraventricularis separated the truncal valve from the tricuspid valve. Mitral-truncal valvular continuity was maintained.

Case 2

Clinical Features. The patient was a 3,175 gm (7 lb) girl. The 42-week gestation of the 23-year-old gravida 2 mother was complicated only by cystitis during the sixth month, for which sulfonamide compounds were administered.

Resuscitation of the infant was not necessary. The Apgar score at one minute was 8 and at five minutes was 10. Slight duskniness of the skin and a grade 2/6 systolic murmur were noted at 30 minutes. A grade 4/6 systolic murmur and a diastolic murmur were noted on the third day after birth. On the fourth day of life, the infant was transferred to the intensive care nursery of St. Mary’s Hospital Medical Center.

Examination on admission at four days of age revealed edema involving the face and eyelids. The infant was mildly cyanotic and somnolent but was easily aroused. The breath sounds were clear with shallow excursions at 60/min. The cardiac rate was 155 beats per minute, and the sounds were accentuated. The first sound was followed by a grade 4/6 systolic murmur and thrill most intense at the fourth left interspace. The murmur was widely transmitted over the anterior wall of the thorax, and a middiastolic flow murmur was heard at the cardiac apex. The second sound was split with a wide interval unchanged during the respiratory cycle. The liver projected 3 cm below the right costal margin. Brachial and femoral arterial pulses were equal and slightly bounding in contour.

A roentgenogram of the thorax revealed cardiomegaly with a globular configuration of the heart and increased pulmonary vascular markings. The ECG showed a short P-R interval, negative delta waves in lead V1, and wide QRS complexes with a duration of 0.10 second, indicating the type B Wolff-Parkinson-White syndrome. This pattern persisted unchanged. The infant was digitalized but remained lethargic, nursed poorly, and continued in a state of congestive failure.

Cardiac catheterization at six days of age (Table 1) re-
FIGURE 4. Systolic pressure of right ventricle measured twice corresponding systolic pressure of truncus arteriosus. High pressure flush (arrow) indicates that truncal pulse recorded through No. 4 French catheter is minimally damped. Electrocardiogram reveals delta waves of preauricular excitation (case 2).

vealed elevated systemic venous pressures and a left-to-right shunt at the atrial level. The right ventricular systolic pressure measured more than twice the aortic systolic pressure (Fig 4). The left ventricle could not be catheterized. Selective cineangiograms revealed large pulmonary arteries which originated from the base of the truncus arteriosus above a narrow root (Fig 5). The truncal valve was narrowed and minimally regurgitant, with only a small amount of contrast medium entering the heart after the truncal injection. A left atroiongram revealed a small left atrium with a large shunt into the dilated right atrium. A small left ventricle and the truncus arteriosus were visualized following flow through the mitral valve. The diagnosis after cardiac catheterization was type 1 or 2 truncus arteriosus with atrial and ventricular septal defects and truncal valvular stenosis.

The infant remained in severe congestive cardiac failure; therapy with digitalis and diuretics was continued. The cardiac size increased progressively, and left pleural effusion developed. When quiet and breathing air, the infant showed nearly normal cutaneous color, but mild cyanosis occurred when she cried. The infant died at 24 days of age.

Pathologic Features. The essential pathologic features were as follows. Arising from the right ventricle was a single arterial vessel. This divided into a short pulmonary trunk, from which the right and left pulmonary arteries and the aorta arose. The aortic arch was left-sided. The truncal valve was quadricuspid and stenotic. The mitral valve was hypoplastic and stenotic. The left ventricular chamber was also hypoplastic, and its only outlet was a narrow muscular-type ventricular septal defect. Thus, there was no mitral-truncal valvular continuity. The chambers of the right atrium and ventricle were enlarged, and the valve of the foramen ovale was herniated toward the right atrium.

DISCUSSION

A diagnosis of persistent truncus arteriosus requires that the systemic, pulmonary, and coronary circulations arise from a common trunk which emerges from the heart. The truncus arteriosus usually arises more from the right ventricle than from the left ventricle. Origin from only the right ventricle is a rare form of the anomaly previously reported by Lampertico (case 43), but without the semilunar valvular stenosis. Dysplasia of the semilunar valves is common in truncus arteriosus.1

FIGURE 5. (case 2). a (left). Catheter entered truncus arteriosus retrogradely from femoral artery and left aortic arch. Dilated pulmonary arteries arise from common vessel. b (right). Dilated and heavily trabeculated right ventricle gives origin to dilated common trunk above narrowed root. Stenotic valve domes into vessel.
Gelband and associates\(^5\) presented autopsy data from 12 babies less than six months of age, seven of whom had deformed and incompetent truncal valves. In these latter seven, refractory cardiac failure had been present.

Van Praagh and Van Praagh\(^6\) reported the pathologic findings in one case with probable truncal valvular stenosis. Victorica and associates\(^7\) reported catheterization data on two patients with significant ventricular-truncal valvular gradients, but without pathologic data. Victorica et al\(^7\) presented phonocardiographic evidence that the second heart sound may be split (as in our case 2) when only one semilunar valve is present. Victorica (personal communication) described the autopsy of another newborn infant with a quadricuspid stenotic truncal valve. Two of the cusps were completely fused. Tandon and associates\(^8\) reported systolic ejection murmurs in eight of 19 cases of persistent truncus arteriosus, but catheterization data revealed no important pressure gradients between either ventricle, on one hand, and the truncus arteriosus, on the other.

As mentioned before, the hemodynamic and pathologic features of our case 1 were similar to the case of truncal valvular stenosis reported by Burnell and associates,\(^2\) with the exceptions that their infant had diminished peripheral pulses, whereas ours did not; and their infant had a left aortic arch, and our case 1 had a right arch. Except for the stenotic semilunar valves, the case of Burnell et al\(^2\) and our case 1 may be considered classic examples of persistent truncus arteriosus in that the common trunk overrode a large ventricular septal defect, and truncal-mitral valvular fibrous continuity was preserved.

Also, as previously mentioned, the hemodynamics and cardiac anatomy of our case 2 had similarities to the case of Lee and associates.\(^3\) The persistent truncus arose only from the right ventricle; therefore, egress from the left ventricle was only through the ventricular septal defect into the right ventricle. Venous pressure and right ventricular systolic and end-diastolic pressures were severely elevated. The main differences were that our case 2 had a stenotic mitral valve, a small left ventricle, and a narrow ventricular septal defect, whereas the case of Lee et al\(^3\) had a large ventricular septal defect and a dilated left ventricle.

Becker and associates\(^1\) performed gross and histologic studies on the semilunar valves from 14 cases of truncus arteriosus. All had abnormal deposits of mucoid connective tissue on the valve cusps, with progression to gross nodularity in nine. The process had extended onto the commissures in four cases. Though none of these valves appeared stenotic, these workers postulated that greater degrees of this dysplastic process might cause stenosis. When the semilunar valve is severely dysplastic in truncus arteriosus, insufficiency is nearly always the only abnormality of hemodynamic importance. Congestive heart failure usually causes the death of these infants by two months of age.\(^5\)

Progression to stenosis can occur, and stenosis was the dominant lesion involving the valves of both babies in our report. Case 1, in whom there was associated insufficiency of the valve, had normal peripheral pulses. The pulses of case 2 were bounding in contour, although the degree of semilunar valvular incompetence was barely appreciable by aortography.

A stenotic semilunar valve may be incompetent to the extent that it cannot close in diastole. A severely stenotic valve may be minimally incompetent, since the orifice is small. The character of peripheral pulses is not entirely dependent on the anatomy of the semilunar valve. In truncus arteriosus, there is free runoff of the blood into the pulmonary arteries, as occurs in the patent ductus arteriosus, and the pulses may have a wide contour even when the truncus valve is competent. Conversely, congestive cardiac failure associated with decreased systemic perfusion causes lowering of the blood pressure and diminishes pulses.

The rapid onset of congestive cardiac failure in our case 1 and the short fulminating course of both patients despite decongestive measures attest to the clinical importance of these truncal valvular lesions. The relatively high oxygen consumption of postnatal life may have contributed to the development of congestive cardiac failure shortly after birth rather than prenatally. The volume overload of a large pulmonary flow was added at birth to the prenatally sustained pressure overload.

We considered surgical palliation of these two infants by valvotomy and banding of the pulmonary arteries. However, it seemed unlikely that integrity of the truncal valve would have been obtained. With the origin of the truncus from the right ventricle in case 2, the stenotic mitral valve and the narrow ventricular septal defect would have obviated a satisfactory operative result.

**References**

The Larynx

Few orators of fame, preachers of great popularity, brilliant actors and actresses of the theater, or outstanding vocal artists of concerts halls and opera houses have been aware or appreciative of the intricate functional marvels of their vocal apparatus. Through eons, phonation has become a universal manner of human communication. It is brought about by the release of puffs of air (air pulses) through the glottis. The number of these puffs, determined by the resilience, tension and vibration of the vocal cords, governs the pitch of the voice when vocalizing. It was Pythagoras (540-500 BC), the Greek philosopher and mathematician, who first ascertained that the pitch of sound is predicated on the length of the vibrating cord; also, he was the discoverer of the mechanism of tonal harmony. In conversational speech the frequency of vibrations of the vocal cords varies from 80 cycles per second in men to 400 cycles per second in women. Throughout recorded history singing has developed into a ubiquitous means for expressing personal joy or sorrow, religious devotion, patriotic dedication, melancholic yearning, and boisterous martial courage. The climax of artistic singing is best exemplified by the phonatory calisthenics of coloratura sopranos, by vocal exhibition in music dramas, and by the bel canto virtuosity of singers in roles identified with melodious, lyrical, romantic arias. The structure of the larynx is based on nine cartilages (three single and three paired): the epiglottis, thyroid, cricoid, arytenoids, corniculates of Santorini and the cuneiforms of Wrisberg. The vocal cords are readily visualized by indirect laryngoscopy. The latter was invented by a singing teacher Manuel Garcia (1805-1906), born in Madrid, teaching in London and Paris. The intrinsic laryngeal muscles modulate phonatory changes of the vocal cords. The functionary role of these muscles is well summarized in one of the medical classic (Gray, H: Anatomy of the Human Body, 29th ed, American ed by Goss, CM Philadelphia, Lea and Febiger, 1974). The posterior cricoarytenoids separate the vocal cords and thus open the glottis. The latter is closed by the lateral cricoytendoids. The unpaired arytenoid closes the glottis, especially at its back part. The cricothyroids tighten and elongate the vocal cords and close the glottis. The main use of the thyroarytenoids is to shorten and relax the vocal cords. "Their lateral portions narrow the riga glottidis by bringing the vocal cords together. Certain minute fibers of the vocalis division, designated as arvyocals muscles are considered by Strong (35) to be chiefly responsible for the control of pitch, through their ability to regulate the length of the vibrating part of the vocal folds." It is truly amazing to witness the facility with which trained singers are able to produce mellifluous, enchanting melodies, or sonorous expression of dramatic emotions by maneuvering the laryngeal structures as if touching the keys of a piano or the strings of a Stradivarius, with virtuoso dexterity. Jackson, C and Jackson, CL (Diseases of the Nose, Throat and Ear, 2nd ed, Philadelphia, Saunders, 1959) offer the following pertinent comments. "The trained soprano, in producing high notes, unconsciously increases the tracheal air pressure coordinately as she firms the cordal edges and shortens the supraglottic air column by raising the larynx and diagnostically diminishes the oral and pharyngeal air column by forcing upward all tissues attached to the hyoid bone. The lips are trained to be kept separated widely. Additionally the trained soprano narrows the ventricle and pharynx." Thus, transposition of lifeless musical notes into beautiful singing is the admirable result of the interplay of the central nervous system, the diaphragm, thoracic and relatively minature muscles of the larynx. The voice of coloratura soprano, Lucrezia Agugari of 18th century Italy, reached up to C in alt, ie a whole octave above high C, corresponding to a vocal cord vibration of 2048 cps as compared to 1408 cps of the vocal cords of other sopranos. What a heavenly revelation it must have been to Mozart who had listened to a recital given by Lucrezia Agugari in Parma, Italy.

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