Truncal Valve Abnormalities in Infants with Persistent Truncus Arteriosus

A Clinicopathologic Study

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SUMMARY
Clinical data on 12 patients under 6 months of age are presented together with the necropsy evidence of truncus arteriosus. Five patients had normal truncal valves and seven had markedly deformed and incompetent valves. Among the group of infants with truncal valve insufficiency, the cause of death was refractory congestive heart failure in each instance, whereas the patients with normal truncal valves succumbed from other causes.

Pathologic examination of the abnormal truncal valves revealed cusps which were thickened by fibrous nodules. Combined heart-lung weight in patients with deformed truncal valves had a mean increase of 85% above normal values, while in the group of patients with normal truncal valves the mean increase was 17%. Similar results were observed when liver weights were compared.

The major clinical findings among the group of patients with truncal insufficiency were bounding peripheral arterial pulses, early diastolic decrescendo murmurs, and congestive heart failure which was refractory to medical treatment and, in one instance, surgical palliation. It is suggested from the data that deformities of the truncal valves are a primary cause of early mortality among infants with truncus arteriosus.

Additional Indexing Words:
Cyanotic congenital heart disease
Pulmonary artery banding
Semilunar valves
Congestive heart failure

TRUNCUS arteriosus (TA) consists of a common arterial trunk supplying the systemic and pulmonary circulations, leaving the heart without a separate pulmonary artery or aorta. The clinical course is variable; many patients die in early infancy with severe congestive heart failure, whereas others survive into older childhood or adult life.1,2 Results of surgical palliation of TA and severe congestive heart failure by pulmonary artery banding have been generally poor.3-5

The explanation for the marked inconsistency in the course of patients with the defect in infancy has not been clear. Although protodiastolic murmurs suggesting truncal insufficiency have been described and anatomic evidence of truncal valvular abnormalities have been reported,6-8 these valvular deformities have not been implicated as being of sufficient importance to be primarily responsible for the early infant mortality. At this institution, clinical experience has suggested...
Table 1

Clinical Data in 12 Infants with Truncus Arteriosus

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Age at death</th>
<th>Type</th>
<th>Cyanosis</th>
<th>Widened pulse pressure</th>
<th>Early diastolic murmur (grade)</th>
<th>Terminal event</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group A</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>M</td>
<td>2 days</td>
<td>I</td>
<td>0</td>
<td>NK</td>
<td>0</td>
<td>Sepsis &amp; pneumonitis</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>4 mo</td>
<td>I</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>Sepsis &amp; meningitis</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>6 mo</td>
<td>I</td>
<td>0</td>
<td>+</td>
<td>0</td>
<td>Pericardial tamponade</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>8 days</td>
<td>II</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>Exploratory surgery</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>3 wk</td>
<td>II</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>Cerebral hemorrhage</td>
</tr>
<tr>
<td>Group B</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>2 wk</td>
<td>I</td>
<td>++</td>
<td>+++</td>
<td>III/VI</td>
<td>CHF</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>3 mo</td>
<td>II</td>
<td>++</td>
<td>+</td>
<td>IV/VI</td>
<td>CHF</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>2 mo</td>
<td>II</td>
<td>+</td>
<td>0</td>
<td>III/VI</td>
<td>CHF; VT</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>1 mo</td>
<td>II</td>
<td>++</td>
<td>+</td>
<td>IV/VI</td>
<td>CHF; VT</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>1 mo</td>
<td>II</td>
<td>+</td>
<td>++</td>
<td>II/VI</td>
<td>CHF</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>2 mo</td>
<td>I</td>
<td>+</td>
<td>++</td>
<td>III/VI</td>
<td>CHF</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>1 mo</td>
<td>II</td>
<td>0</td>
<td>+++</td>
<td>III/VI</td>
<td>CHF</td>
</tr>
</tbody>
</table>

Abbreviations and symbols: 0 = none; + = mild; ++ = moderate; +++ = marked; CHF = congestive heart failure; VT = ventricular tachyarrhythmias; NK = not known.

that severe malformations of truncal valves are an important causative factor in early deaths among infants with this anomaly. Accordingly, autopsies of patients with TA who died before the age of 6 months at Columbia Presbyterian Medical Center were reviewed to correlate the clinical course and cause of death with the anatomy of the truncal valves.

Clinical Data

A total of 12 patients with TA who were less than 6 months of age came to postmortem examination at Babies Hospital. All were classified as having TA of type I or II according to Collett and Edwards. Gross and microscopic examinations were available in each instance. Five of the infants had normal truncal valves (group A), while seven (group B) were found to have marked gross and microscopic valvular abnormalities.

Pertinent clinical information is presented in table 1. The majority of the infants were under 2 months of age at the time of death. Cyanosis was absent or mild in the five patients in group A. In group B, one infant was observed to be acyanotic, whereas the remaining six had mild-to-moderate degrees of clinical cyanosis. Peripheral pulses were normal or mildly increased in prominence among infants in group A, whereas five patients in group B displayed strikingly bounding pulses consistent with a widened pulse pressure by palpation. One infant was described as having normal pulses.

Each of the 12 patients had a holosystolic murmur of varying intensity that was audible along the left sternal border. No infant in group A had an early diastolic murmur audible along the left sternal border, but two patients had rumbling middiastolic murmurs at the apex suggestive of increased mitral flow. In contrast, all patients in group B had early diastolic blowing murmurs along the midleft sternal border, typical of aortic insufficiency. Continuous murmurs were not described in either group.

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Electrocardiographic and X-Ray Data

There were no significant differences in the electrocardiograms obtained from patients in group A or B. Tall peaked P waves (2.5 mm) diagnostic of right atrial enlargement were present in three cases. The mean QRS axes in the frontal plane were between +60° and +120°. Two patients with QRS axes of +60° had evidence of biventricular hypertrophy, but the majority of the electrocardiograms showed evidence of right ventricular preponderance which were considered to be within normal limits for age.

All patients had X-ray evidence of cardiomegaly with pulmonary vascular engorgement ranging from moderate to severe.

Clinical Course

The clinical course and cause of death were dissimilar in these two groups. Patients in group A responded well to digitalis and diuretics during early congestive heart failure, and none had evidence of cardiac decompensation at the time of death. Two died of sepsis, confirmed by positive blood and cerebrospinal fluid cultures; a third succumbed from a massive cerebral hemorrhage. Another infant suffered a ventricular perforation during cardiac catheterization carried out in 1959, with resultant pericardial tamponade; and the fifth baby underwent elective exploratory surgery in 1952 with the erroneous diagnosis of a patent ductus arteriosus and died immediately following operation. The cause of death in these five infants was not directly related in their cardiac lesion.

In contrast, none of the patients in group B responded to maximal cardiotonic regimens, and all died with refractory congestive heart failure. Two of the infants succumbed with terminal ventricular tachyarrhythmias.

Surgery

One patient in group B (case 6), underwent pulmonary artery banding at 13 days of age. Cardiac decompensation did not improve after the attempt at surgical palliation, and the infant died 36 hours after operation with unremitting heart failure. Shortly before death, the most striking clinical feature was the bounding peripheral pulses. The dorsalis pedis pulse was not only palpable but could be easily visualized.

Pathologic Findings

The hearts were enlarged with markedly thickened ventricular musculature. All of the patients in both groups had ventricular septal defects, one had an associated atrial septal defect, and one a persistent left superior vena cava.

No significant noncardiac anomalies were present in either group of infants.

The truncal semilunar valves of the group A patients had three cusps and were grossly and microscopically normal. In group B, six of the patients had three truncal valve cusps, two with partial raphes, and the seventh had four cusps. All of the valves were thickened and opaque with malformed nodular margins, and in each instance close apposition of the cusps was impossible. The gross anatomy of normal (case 3) and abnormal (case 6) truncal valves is shown in figure 1A and B, and the sagittal microscopic views from the same patients are shown in figure 2A and B. As noted in figure 2B, microscopic sections of the abnormal valve demonstrated marked proliferation of loose fibrous tissue, with nodules of fibrinoid and collagen material.

There was gross and microscopic evidence of passive congestion of the lungs and liver in all the patients, but this finding was more striking among the infants in group B. To assess organ congestion, the combined heart-lung, which had been fixed intact, and liver weights were compared to known normal values. Although all group A patients had heart-lung and liver weights greater than normal, none exceeded 1 standard deviation increase above the normal mean weights for age, whereas all patients in group B had organ weights greater than 2 standard deviations above normal. The heart and lung weight increased 8 to 32% in patients in group A (mean, 17%) compared to 71 to 98% in group B (mean, 85%). Liver weights had an increase of 4 to 12% above normal among infants in group A (mean, 8%), while a more
striking mean increase of 52 to 84% was found in patients in group B (mean, 67%). Thus, heart-lung and liver weights correlated well with the clinical diagnosis of unremitting congestive heart failure in group B patients.

**Discussion**

Gross and microscopic abnormalities of the semilunar valves in truncus arteriosus (TA) were initially described by Motta in 1932. Three years later, Roos fully described the pathology of abnormal truncal valve cusps in a 2-week-old neonate with the defect. More recently, Deely and associates observed truncal valve abnormalities in five infants less than 1 month of age in a postmortem study of 43 cases of TA of all ages, and McNamara and Sommerville reported a diastolic murmur diagnostic of truncal valve incompetence in nine of 50 cases.

The specific etiology of truncal valve deformities is unknown. Microscopic examinations of the abnormal valve cusps among patients in this report as well as other studies have demonstrated nonspecific fibrous proliferation in the malformed nodular margins of the valves. Whether these nodular fibrous proliferations are secondary to inflammatory changes, or whether they represent a facet of the congenital defect among some cases of TA, is open to conjecture and cannot be ascertained from routine pathologic study. Roos' etiologic concept.

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**Figure 1**

(A) Gross postmortem specimen of patient 3 in group A. Note right ventricle (RV), high ventricular septal defect (VSD), overriding common truncus (CT), and small arrow pointing to orifice of main pulmonary artery (MPA). The truncal valve (TV) cusps are smooth and normally formed.

(B) Gross postmortem specimen in case 6 in group B. The orifice to the main pulmonary artery (MPA) is visualized arising from the common outflow trunk (CT). The truncal valves (large arrow) are thickened, nodular, and deformed.

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In none of the reports of truncal valvular deformities in TA^6-8 were clinical, pathologic, or prognostic correlations made according to the presence or absence of truncal valve deformities. Congestive heart failure was observed among infants whether or not truncal insufficiency was present. The left ventricle, in TA, may decompensate as a result of an increased volume load secondary to increased pulmonary flow and left atrial return. However, the additional burden of truncal valve insufficiency increases heart failure, which leads to decreased forward cardiac output and pulmonary blood flow. This results in lower peripheral arterial O₂ saturation. The preceding sequence of events would account for the refractory congestive heart failure and more prominent cyanosis observed clinically in the group of infants with truncal valve abnormalities.

In the light of the grave prognostic implications of truncal valve incompetence, it becomes of great importance to focus attention on the early identification of truncal valve incompetence in babies with TA. Table 2 presents the major clinical manifestations in

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Figure 2

(A) Saggital microscopic view taken from specimen in figure 1A showing a normal truncal valve (TV) which is smooth, thin, and free of nodules and convolutions. Atria (A) and ventricles (V) are marked for orientation.

(B) Microscopic saggital view of abnormal truncal valve (TV) taken from heart in figure 1B. Note marked convolutions and thickened nodular appearance of the valve.
patients with normal truncal valves and those with severe truncal insufficiency. In general, clinical differentiation should not be difficult. Bounding peripheral pulses and an early diastolic decrescendo murmur are characteristically observed with truncal insufficiency, whereas patients with normal truncal valves usually have normal or slightly increased peripheral pulses and no diastolic murmur suggestive of semilunar insufficiency. However, in order to delineate clearly the anatomy of the truncal valves, cardiac catheterization and angiography is mandatory. An angiogram of the ascending aorta, preferably made by the retrograde route, will not only determine the details of the origin of the pulmonary arteries from the truncus, but will also confirm the presence or absence of associated truncal valve incompetence.

The presence of truncal insufficiency is also an important factor in the prognosis for surgical palliation in infants with TA. A higher mortality has been reported after pulmonary artery banding in patients with this defect than with several other forms of congenital heart disease.3-5 Part of the explanation for the poor results may be that the surgical procedure may not ameliorate the major hemodynamic abnormality responsible for the intractable left ventricular decompensation (that is, truncal valvular insufficiency). This was true in the infant (case 6) described in this report in whom the congestive heart failure continued unabated after operation. Bounding peripheral pulses and a prominent early diastolic murmur, in the presence of a low cardiac output state, strongly suggest that the major factor in this patient's death was truncal valve incompetence.

An early definitive operative approach to malformed truncal valve awaits further advances in infant valve surgery. Thus, despite the poor results of pulmonary artery banding in TA, at present, little else is available surgically for these infants. However, vigorous efforts should be made at medical or surgical palliation since definitive correction may be possible at a later age. At this institution and elsewhere older children and adults are alive despite truncal valve abnormalities with resultant valvular incompetence and some have even undergone total surgical correction. McGoon and associates13 reported no significant relationship between the risk of mortality and the preoperative presence of truncal valve insufficiency in the total repair of TA. In all likelihood, the child who has survived the neonatal period, with time, is able to compensate for the additional volume work of truncal insufficiency and become a candidate for corrective surgery.

In most series, marked truncal valve incompetence in older infants and young children with TA is not described as a serious problem,1-2 suggesting that patients with the most severe truncal valve abnormalities are eliminated at a younger age by natural selection. The clear definition of the grave clinical profile among infants with truncal valve deformities presented here is a further indication that truncal insufficiency is a major negative prognostic factor in the first months of life. This fact should be a prime consideration in the diagnostic evaluation and surgical management of these patients.

Table 2

Clinical Comparison of Infants with Normal Truncal Values to those with Truncal Insufficiency

<table>
<thead>
<tr>
<th>Competent truncal valve</th>
<th>Truncal valve insufficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arterial pulse</td>
<td>Normal to slightly full</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>Variable</td>
</tr>
<tr>
<td>Early diastolic murmur</td>
<td>Absent</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>Responds to medical therapy</td>
</tr>
<tr>
<td>Course</td>
<td>Often survives infancy</td>
</tr>
<tr>
<td></td>
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</table>
TRUNCAL VALVE ABNORMALITIES

References

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