Truncus Arteriosus
Clinical Study of Fourteen Cases

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We have encountered difficulty in recognizing and identifying clinically true truncus arteriosus with pulmonary arteries (type 1) because of its resemblance to correctable cardiac defects such as patent ductus arteriosus, aortic-pulmonic defect, and ventricular septal defect. In the present report we are presenting clinical information on 14 patients with this anomaly, both to supplement the scanty data available in the literature and to emphasize the need for correct diagnosis.

Persistent truncus arteriosus is defined as a single trunk arising from the heart and supplying the coronary, pulmonary, and systemic circulations, with no remnants of an atretic aorta or pulmonary artery.1-4 The number of semilunar cusps may vary from 2 to 6. Typically a defect is present in the bulbar portion of the anterior ventricular septum, a defect lying just under the semilunar cusps and entirely different from the usual ventricular septal defect involving or bordering on the membranous portion of the septum. Photographs of an autopsy specimen (case 10) are shown in figures 1 and 2.

Considerable disagreement has arisen in the classification of truncus defects, particularly with regard to the inclusion of cases having only bronchial circulation to the lungs. Mora-gues4 preferred to exclude such cases, whereas Collet and Edwards3 included them as a special subtype. Kjellberg and associates5 included them with the "pseudotruncus" group because of the great clinical similarity. Manhoff and Howe6 included truncus arteriosus in their classification of defects involving absent or anomalous pulmonary arteries. MacGilpin7 separated cases with 1 pulmonary artery from those with 2. Cases have also been classified as to the origin of the trunk, whether from the right ventricle or left ventricle or "overriding," or whether a single ventricle is present.8

The following simplified classification is modified from those of previous workers. This classification not only follows natural embryologic and anatomic divisions, but also appears to be practical from a clinical viewpoint. Whether the pulmonary arteries arise separately but in very close approximation, or whether the pulmonary artery arises singly and then divides into 2 main branches, is often difficult to decide in individual cases, even at the autopsy table. Since such cases are closely related embryologically and clinically, it appeared logical to combine those cases previously called type 1, type 2, and type 3 by Collett and Edwards.9 On the other hand, the absence of 1 pulmonary artery changes the clinical picture considerably, and to us merits separate classification.

In view of the considerable differences of opinion regarding classification, it seems advisable for individual cases to be referred to in descriptive terms (true truncus arteriosus with pulmonary arteries, true truncus arteriosus with absent pulmonary arteries, etc.) rather than in arbitrary terms (type 1, type 2, etc.). Unfortunately, there are some intermediate cases, with relatively small pulmonary arteries and large bronchial arteries (case 6 in present series). Likewise, as will be discussed later, cases of our type 1 truncus arteriosus do not approach a stereotyped clinical pattern, and associated defects may modify the picture even further.

Classification of Truncus Malformations

True Truncus Arteriosus

Type 1. True truncus arteriosus, with pulmonary arteries arising from the trunk proximal...
to the innominate artery (includes types 1–3 of Collett and Edwards3).

**Type 2.** True truncus arteriosus, with absence of 1 pulmonary artery.

**Type 3.** True truncus arteriosus, with absence of both pulmonary arteries.

**Type 4.** Partial truncus arteriosus (aortopulmonary defect or “window”).

“Pseudotruncus”

**Type 1.** Solitary aortic trunk with pulmonary atresia.

**Type 2.** Solitary pulmonic trunk with aortic atresia.

The embryologic features of persistent truncus arteriosus, as well as the case literature, have been carefully reviewed by Humphreys,6 Lev and Saphir,2 Collett and Edwards,3 and MacGillip.7 On the other hand, relatively little has been written regarding the clinical picture presented by patients with this defect. We have studied 14 cases of true truncus arteriosus having 2 pulmonary arteries (type 1) and have been impressed by the wide range and confusing nature of the clinical findings. These 14 cases have all been verified by surgical exploration or postmortem examination. We have studied an additional half dozen cases in which persistent truncus arteriosus of similar type has been diagnosed clinically, but which are being omitted in view of the lack of direct confirmation.

The clinical and laboratory data on these 14 cases are summarized in tables 1 to 5. In addition, 1 case is being reported in detail to illustrate the difficulties in clinical diagnosis.

**Case Report**

**Case 3** (R.O.). This boy was referred to University Hospitals on August 31, 1950, at the age of 4 years. The mother reported that she had had a 2-day attack of mild measles (rubella?) during the first trimester of pregnancy, and that a daughter had had a similar illness at the same time. A heart murmur had been first heard when the boy was 2 months of age. During his first year of life he gained poorly and suffered 2 attacks of pneumonia. Subsequently he did well, except for slight dyspnea on exertion.

On examination, the boy appeared small for his age, weighing only 31 pounds and measuring 38 inches in height. No cyanosis was noted. A precordial bulge was present. There was a soft systolic thrill and short harsh systolic murmur along the lower left sternal border. The pulmonic second sound was not
accentuated and was followed by a long blowing diastolic murmur. Blood pressure was 90/60 in the right arm. The hemoglobin concentration was 12.5 Gm./100 ml. and the hematocrit value was 37 per cent. The electrocardiogram (no precordial leads) showed an axis of +100 degrees and notching of the P waves in lead I. Roentgenography showed moderate cardiac enlargement, involving both ventricles, with increased pulmonary vascular markings.

A diagnosis of interatrial septal defect was considered most likely. Cardiac catheterization was then performed, and an increase in oxygen content was found in the "main pulmonary artery" as compared to the right ventricle. Angiocardiography at this
time showed normal filling of the pulmonary vessels, with a suggestion of minimal opacification of the descending aorta. On the basis of these findings a diagnosis of aortic-pulmonic "window" was considered most likely.

The boy was next seen 2 years later (1953) and was then reported to be getting along well. No cyanosis was noted at that time. A precordial bulge and systolic and diastolic murmurs were again noted. Blood pressure was 84/50 in the right arm. The preferred diagnosis was now Eisenmenger complex, a diagnosis made meanwhile at another cardiac clinic.

In 1954 his case was reviewed and the possibility of a reversing patent ductus arteriosus was considered. Simultaneous blood samples were obtained from the right brachial and femoral arteries, but both showed equal degrees of desaturation. He was readmitted for repeat cardiac catheterization in September 1954. At this time he was still reported to have only mild exertional dyspnea. He appeared small and showed no cyanosis. Cardiac findings were essentially unchanged, except that mention was now made of accentuation of the pulmonic second sound. The hemoglobin concentration was 15.0 Gm./100 ml. and the hematocrit level was 44 per cent. The electrocardiogram showed an axis of +110 degrees notched P waves in lead I, and evidence of right ventricular preponderance on the precordial leads (R/S in V1 was 14/2; in V6, 16/35). Roentgenography again showed considerable cardiac enlargement, with marked prominence of the "pulmonary artery segment" and pulmonary vasculature; the aortic arch was on the right (fig. 3). At cardiac catheterization the catheter tip was passed into the ascending and descending aorta, but the pulmonary arteries could not be entered. The findings were considered to indicate a ventricular septal defect with bidirectional shunt.

Cardiac surgery was scheduled for October 8, 1954, with the use of the cross-circulation technic. On opening the pericardium the surgeons identified a truncus arteriosus defect. The main vessel measured 7.5 cm. in diameter. Approximately 3 cm. above the origin of this trunk there arose a pulmonary artery, 3 cm. in diameter, which then divided into left and right branches. No attempt at correction was made. Convalescence was without complications and the patient was discharged on the eleventh postoperative day. He was next seen in April 1955, at which time he was reported to be getting along very

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**Table 3.—Roentgenographic Findings**

<table>
<thead>
<tr>
<th>Case</th>
<th>Heart size</th>
<th>Chamber enlargement</th>
<th>Upper left border</th>
<th>Pulmonary vascular markings</th>
<th>Aortic arch</th>
<th>Miscellaneous</th>
<th>Angiocardiogram</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. C. P.</td>
<td>4+</td>
<td>4+ 4+ N</td>
<td>Marked bulge</td>
<td>3+</td>
<td>L-large</td>
<td>Emphysema</td>
<td>—</td>
</tr>
<tr>
<td>2. D. P.</td>
<td>2+</td>
<td>2+ 2+ 1+</td>
<td>Convexity</td>
<td>2+</td>
<td>L</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>3. R. O.</td>
<td>2+</td>
<td>2+ 2+ N</td>
<td>Convexity</td>
<td>3+</td>
<td>R-large</td>
<td>Heart shifted to right</td>
<td>No early filling of aorta</td>
</tr>
<tr>
<td>4. L. K.</td>
<td>2+</td>
<td>3+ 1+ N</td>
<td>Slight convexity</td>
<td>1+</td>
<td>L-large</td>
<td>—</td>
<td>Right to left shunt in atrium; simultaneous filling of PA and aorta</td>
</tr>
<tr>
<td>5. V. E.</td>
<td>3+</td>
<td>3+ 3+ 2+</td>
<td>Flat</td>
<td>2+</td>
<td>R</td>
<td>—</td>
<td>No early filling of aorta</td>
</tr>
<tr>
<td>6. M. H.</td>
<td>3+</td>
<td>3+ 1+ 1+</td>
<td>Flat</td>
<td>2+</td>
<td>R</td>
<td>—</td>
<td>RPA filled from trunk, LPA from descending aorta. Aortogram: RPA filled from R ductus, LPA from descending aorta</td>
</tr>
<tr>
<td>7. D. R.</td>
<td>3+</td>
<td>2+ 4+ 2+</td>
<td>Flat</td>
<td>3+</td>
<td>L</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>8. M. M.</td>
<td>—</td>
<td>— — —</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>9. H. W.</td>
<td>3+</td>
<td>2+ 2+ 2+</td>
<td>Flat</td>
<td>3+</td>
<td>L-large</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>10. J. G.</td>
<td>3+</td>
<td>2+ 2+ N</td>
<td>—</td>
<td>2+</td>
<td>L</td>
<td>Ledge in LAO</td>
<td>—</td>
</tr>
<tr>
<td>11. P. W.</td>
<td>3+</td>
<td>3+ 3+ 2+</td>
<td>Slight convexity</td>
<td>3+</td>
<td>L</td>
<td>—</td>
<td>Inadequate</td>
</tr>
<tr>
<td>12. T. W.</td>
<td>3+</td>
<td>3+ 1+ 1+</td>
<td>Marked convexity</td>
<td>3+</td>
<td>L</td>
<td>—</td>
<td>Simultaneous filling of PA and aorta</td>
</tr>
<tr>
<td>13. D. L.</td>
<td>3+</td>
<td>— — —</td>
<td>—</td>
<td>2+</td>
<td>L</td>
<td>Heart in right chest</td>
<td>—</td>
</tr>
<tr>
<td>14. C. T.</td>
<td>1+</td>
<td>N 1+ N</td>
<td>Slight convexity</td>
<td>2+</td>
<td>R</td>
<td>—</td>
<td>No early filling of aorta</td>
</tr>
</tbody>
</table>
TRUNCUS ARTERIOSUS

Table 4.—Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Location*</th>
<th>Case 3†</th>
<th>Case 4</th>
<th>Case 9†</th>
<th>Case 14‡</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1/12/51</td>
<td>9/29/54</td>
<td>1/28/55</td>
<td>2/1/55</td>
</tr>
<tr>
<td>SVC</td>
<td>11.1</td>
<td>12.4</td>
<td>6.2</td>
<td>5.9</td>
</tr>
<tr>
<td>IVC</td>
<td>9.4</td>
<td>11.1</td>
<td>2.6</td>
<td>—</td>
</tr>
<tr>
<td>RA</td>
<td>10.7</td>
<td>11.6</td>
<td>6.4</td>
<td>6.6</td>
</tr>
<tr>
<td>RV</td>
<td>12.2</td>
<td>14.3</td>
<td>4.0</td>
<td>7.0</td>
</tr>
<tr>
<td>“PA”</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>“MPA”</td>
<td>14.4</td>
<td>—</td>
<td>8.3</td>
<td>—</td>
</tr>
<tr>
<td>“RPA”</td>
<td>14.8</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Car Art</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>R Br Art</td>
<td>—</td>
<td>16.7</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>L Br Art</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>9.4</td>
</tr>
<tr>
<td>Aorta</td>
<td>—</td>
<td>16.5</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Fem Art</td>
<td>14.9</td>
<td>—</td>
<td>8.1</td>
<td>7.5</td>
</tr>
</tbody>
</table>

*Quotation marks are used to indicate doubt as to correct identification of location, though they were called this at the time of cardiac catheterization.

† Simultaneously drawn samples of blood (8/19/54) from right brachial artery and femoral artery showed equal degrees of oxygen desaturation (88 per cent).

‡ Two sets of data obtained at different times during same cardiac catheterization. Repeat femoral artery oxygen determination on 11/8/55 showed 92.8 per cent saturation.

§ Samples from aorta in second cardiac catheterization were taken as catheter tip was slowly withdrawn from descending aorta (14.3) to “ascending aorta” just above valve (12.9).

well, with little or no cardiac symptoms. His weight was only 49 pounds (age 8 years), and height, 48 inches. The cardiac findings were unchanged.

Additional Data

Various data not included in the tables are also of interest. Among the 13 cases in which detailed pregnancy histories were obtained, there was a history of “mild measles” in one (case 3), inadequate diet in another (case 9), and polyhydramnios in a third (case 6). Birth weights showed a normal average of slightly over 7 pounds, though in 1 case the birth weight was only 4½ pounds. Twelve of the 14 births occurred between June 15 and December 15. There were 10 males and 4 females. None of the patients was a member of a twin pair. There were 13 older living siblings; of these, 2 had cyanotic congenital heart defects. One
older sibling had died at birth "with the cord around his neck." There were only 2 younger siblings, both apparently normal. Case 6 was of considerable interest, since the mother has had 4 children by 4 different men; the oldest child has cerebral palsy and mental retardation, the next has cyanotic congenital heart disease (probably severe tetralogy of Fallot), the next is case 6, and the fourth, just recently born, is said to be normal. The foregoing data are too limited in themselves to allow for any conclusions, but if combined with data of others, may be of value. Whether the seasonal incidence mentioned above is of etiologic significance, as in patent ductus arteriosus, remains to be seen.

**Discussion**

**General Features**

As already mentioned, our 14 cases presented a wide variety of clinical findings. The majority died in early infancy, but 3 lived to at least 7 years of age. Abbott1 tabulated 21 cases with death occurring from birth to 25 years, the average being 4 years. In his review of the literature MacGilpin7 cited cases living up to the age of 36 years, but 85 per cent died before 2 years of age. Incidentally, cases of truncus arteriosus without pulmonary arteries showed a slightly better life expectancy in both of these series of cases. In both there was a slight excess of males.

Growth retardation was a typical feature of all of our cases, even in those dying in infancy. Respiratory infections tended to be frequent and severe, though in 2 of our oldest cases this did not represent a problem after the first 2 years of life. Dyspnea developed sooner or later in all, but in our cases did not necessarily parallel the level of cyanosis. The patient may die in either left or right heart failure.

**Physical Findings (table 1)**

Other writers10-12 have described cyanosis (some pointing out that it may be minimal or even absent) and a loud parasternal systolic murmur, with occasionally a machinery-type
murmur, as characteristic findings. Contro and associates reported 4 cases mimicking patent ductus arteriosus, in 3 of which there was a rumbling murmur extending through systole into but not through diastole; however, the murmur was not considered to be of typical ductus quality, nor was it the "familiar soft continuous bruit commonly heard in patients with truncus and dilated bronchial arteries." Our cases showed extreme variations in cyanosis, from absent to severe, even in those of equal age. Loud systolic murmurs along the left sternal border were present in all of our cases, usually with an associated thrill; but a diastolic murmur was heard only in the 3 oldest cases, where it was present over the pulmonic area. In none was there a machinery-type murmur. Apparently a true machinery-type murmur is rare or exceptional in type I truncus arteriosus defects, and the widespread impression that it does occur has probably arisen from the observations of continuous systolic-diastolic bruits in type 3 truncus arteriosus defects (absent pulmonary arteries, pulmonary circulation being by way of large bronchial arteries).

Taussig described a loud pure second sound over the base as a specific feature of this defect, a sign emphasized also by others. Unfortunately, in our series of cases, with the exception of the 3 most recent cases, examiners failed to comment on the nature of the second sound except to mention its intensity. However, no mention was made in any case of a split second sound over the base. On the other hand, Gotzsche and Singleton's group have both reported clinically diagnosed cases of truncus arteriosus with split second sounds. Neither of these cases has been verified by direct observation. It is possible that other heart sounds could mimic a splitting, but it is difficult to reconcile a true splitting of the second sound with this defect. Phonocardiography might well prove to be extremely useful in evaluating the nature of the second heart sound when a truncus arteriosus defect is suspected.

Blood pressures showed no consistent pattern as far as pulse pressure was concerned and were generally within normal limits. A precordial bulge appeared to be one of the more characteristic findings.

Electrocardiographic Findings (table 2)

As with other clinical features, the electrocardiogram in our cases showed considerable variation. Usually there was slight right axis deviation, a finding normal in the age range studied. Peaking of the P waves in lead II appeared to be the only frequent abnormal finding. Of the 2 older children with minimal arterial oxygen desaturation, one had right ventricular preponderance and the other had left ventricular preponderance as well as left ventricular strain. In general there is little of diagnostic significance in the electrocardiogram, a comment already made by Taussig. Others have mentioned right axis deviation or either right, left, or combined ventricular hyper trophy.

Roentgenographic Findings (table 3)

Taussig described the typical roentgenographic findings of truncus arteriosus, including type I and type 3, in infancy as cardiac enlargement, pulmonary concavity, upturned apex, a prominent aortic knob, and a "shelf" in the left anterior oblique view. In older patients the heart was said to have a contour similar to a tetralogy of Fallot with severe pulmonary stenosis. If the lungs were supplied only by bronchial arteries (type 3 in our classification), then there was said to be diminished hilar shadows. Gasul and co-workers have also emphasized pulmonary concavity, upturned apex, and prominent aortic knob. Abrams has described a high "hilar comma" and the frequent occurrence of a right aortic arch in this defect.

In our cases roentgenography has typically shown moderate to marked cardiac enlargement, with right as well as some left ventricular enlargement, and a moderate increase in pulmonary vascular markings. The left atrium was enlarged in about half the cases. No typical cardiac contour was found, but an oval or egg-shaped outline, as described by Rowe and Vlad in a case with a right aortic arch, was seen several times. We encountered it however in cases with left aortic arch but in none of the 4 cases with right aortic arch. The "pulmonary artery segment" (upper left border) was generally flat, but in several instances showed a
prominent though rather high bulge that actually represented the left pulmonary artery; this bulge could be identified as being separate from the heart border in this area sometimes on the plain films and sometimes on fluoroscopy. Variations in cardiac contour are illustrated in figures 3 to 6.

In only a minority of cases was the “ascending aorta” or “aortic knob” noted to be very large. A “shelf” in the left oblique view was seen in only 1 case; incidentally, this “shelf” is frequently seen in cases of true truncus arteriosus with absent pulmonary arteries (type 3 in our classification) or in “pseudotruncus.” We found a high “hilar comma” to be present in about half of our cases, particularly in the older patients.

Since the main pulmonary artery segment is absent anatomically in true truncus arteriosus, careful film and fluoroscopic observation should be made to determine its absence. In infants, observation is made difficult by the thymus gland, which so often obscures the region of the main pulmonary artery segment. In the presence of increased pulmonary vascular flow, transposition of the great vessels and its subtypes (transposition with tricuspid atresia, corrected transposition, and single ventricle with transposition and rudimentary outflow for the aorta) are the only other conditions that also have absent main pulmonary artery segments on roentgen examination. Therefore, if the absence of the main pulmonary artery segment can be determined, the finding becomes im-
portant in that it immediately excludes a number of conditions. Coupled with the other findings of high “hilar comma,” active and often prominent “aorta,” and increased pulmonary vascularity, the absence of the main pulmonary artery segment should direct one’s attention to the possibility of a true truncus arteriosus defect.

Findings on Angiocardiography and Aortography (table 3)

Angiocardiography has been generally used or described as a means of clinically identifying truncus arteriosus, though apparently no one individual or group has had extensive experience with this type of defect. Nevertheless, Abrams believed it to be the only method of establishing a conclusive diagnosis of truncus arteriosus. He stated that in this defect there is immediate filling of the trunk from the right ventricle, with no contrast material in the pulmonary artery until the trunk is filled; the trunk is said to be twice the ordinary size of an ascending aorta, and becomes much smaller beyond the isthmus; the left ventricle often fills through the ventricular septal defect. None of the above authors referred to failures or to the difficulties that might be expected in cases where the peripheral arterial oxygen saturation approaches normal values. However, DeGroot mentioned 1 failure, and recommended that angiograms be obtained in both the anteroposterior and left oblique positions.

Our experience with angiography in cases of truncus arteriosus is limited. This procedure was done in 7 of the patients here reported. We have not had the benefit of a biplane camera, a very limiting factor in our opinion. In several of our cases, technical difficulties resulted in nondiagnostic films. Angiocardiography permitted a positive diagnosis in cases 6 and 12. Both of these patients were cyanotic, and consequently simultaneous opacification of the trunk, pulmonary arteries, and aorta would be expected. On the other hand, in cases 3 and 14, who were only minimally desaturated, the aorta did not opacify sufficiently for early radiologic detection.

Aortography as a means of diagnosing truncus arteriosus defects has been used very little, or at least little has been published on this subject. In describing their experience with aortography in infants, Keith and Forsyth include 1 case of truncus arteriosus that was erroneously diagnosed by aortography as a patent ductus arteriosus. With reference to such confusion, they write, “It may be difficult at times to eliminate the possibility of a persistent truncus by this method, but there are certain features that help to differentiate the two conditions. When the ductus fills from the pulmonary artery, the latter usually shows as a large vessel close to the aorta. On the other hand, the vessels arising from the aorta, in persistent truncus, are apt to be smaller than the main pulmonary artery. In persistent truncus, one may detect that the vessels going to the lung arise from the ascending aorta and in this way be able to distinguish them from the ductus which arises from the descending aorta.” Singleton and associates found aortography of ancillary help in 1 of their cases. Abrams also has mentioned possible confusion of truncus arteriosus with patent ductus arteriosus on aortography, but apparently did not consider this procedure particularly useful in diagnosing the former. This confusion is further emphasized by Contro and associates, who reported 4 cases of clinically diagnosed “atypical” patent ductus arteriosus, with retrograde arteriograms showing opacification of the pulmonary vessels from the aorta, but which at surgery proved to be truncus arteriosus defects.

Only 1 of our cases had aortography, and in this instance (case 6) it was of some diagnostic help. If aortography is to be performed, it would appear ideal to do the injection through a catheter, with the tip of the catheter in the “ascending arch.” One would expect the force of injection to alter locally the hemodynamics of blood flow, and thereby disrupt the “streaming” patterns in the single trunk and allow for simultaneous opacification of the pulmonary arteries and aorta. This would be particularly useful in those cases where the systemic arterial oxygen saturation approaches normal. To be sure, there would be some confusion with a partial truncus (aortic-pulmonary defect), and other diagnostic information would be necessary.
Cardiac Catheterization Findings (table 4)

Our limited experience with cardiac catheterization in this defect does not allow for broad generalizations. Actually, we were misled several times by misinterpretation of the data presented in table 4. It is obvious that pressures in the right ventricle will approximate those in the left. Likewise, because of the ventricular septal defect, the oxygen content in the right ventricle is usually increased as compared to the right atrium. Moreover, a further increase in oxygen content may be expected in the pulmonary arteries, coupled with systemic arterial oxygen desaturation. The oxygen saturations of the pulmonary arteries and systemic arteries may well differ, due to differential streaming. The systemic arterial oxygen saturation may actually be within "normal limits." Considerable difficulty arises in failure to identify correctly the location of the catheter tip when arterial areas are entered. The truncus can usually be entered, but its anomalous location and direction may be confusing if the defect is not suspected ahead of time. There is greater difficulty in entering the pulmonary arteries than in passing the catheter tip up into the aortic arch. In several cases diagnosed clinically but not verified by direct observation we have entered an arterial trunk from an anomalous position and have failed to enter the pulmonary arteries.

Confusion from catheterization, particularly in the cases with borderline systemic arterial oxygen saturation, centers around the exclusion of aortic-pulmonic defect, patent ductus arteriosus, or ventricular septal defect with marked pulmonary hypertension. Dheer and van Nieuwenhuizen have described a method of identifying the first of these by passage of the catheter through the defect and then down toward the aortic valve, as well as upward into the arch of the aorta, with films being taken of the catheter in these positions. It is true that a significant increase in oxygen content in the right ventricle would tend to rule out an aortic-pulmonic defect. A nonreversing patent ductus arteriosus can be identified if the catheter is passed through it in typical fashion. A reversing patent ductus arteriosus can be identified by the difference in oxygen saturations of the right brachial and femoral arteries. An isolated ventricular septal defect can be ruled out if the catheter can be manipulated from the aortic arch to a pulmonary artery without a change from an arterial type of pressure tracing. The limiting factor in all of these situations is of course the frequent inability to direct the catheter tip into all of the desired structures. Dye-dilution technics might well be expected to supplement cardiac catheterization as a means of detecting right-to-left shunts in those cases where the shunt is too small to be seen on angiocardiography. Some of the foregoing clinical features are listed in table 6 for use in differential diagnosis. The presence of combined defects, such as patent ductus arteriosus with ventricular septal defect, increases the chances for diagnostic error.

Autopsy and Surgical Findings (table 5)

All but 2 of the autopsied cases had a truncus guarded by 3 semilunar cusps. The 2 exceptions

<table>
<thead>
<tr>
<th>Defect</th>
<th>Second heart sound</th>
<th>Size of &quot;ascending aorta&quot;</th>
<th>Cardiac catheter passes into &quot;Ascending aorta&quot; and carotid artery</th>
<th>Increase in oxygen content occurs in</th>
</tr>
</thead>
<tbody>
<tr>
<td>Truncus arteriosus</td>
<td>Always pure</td>
<td>Large</td>
<td>Usually</td>
<td>Seldom</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>Usually split</td>
<td>Large</td>
<td>Never</td>
<td>Almost always</td>
</tr>
<tr>
<td>Aortie-pulmonic defect</td>
<td>Usually split</td>
<td>Normal to large</td>
<td>Often</td>
<td>Almost always</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>Usually split</td>
<td>Small</td>
<td>Occasionally</td>
<td>Almost always</td>
</tr>
</tbody>
</table>

TABLE 6.—Differential Diagnosis for a Patient with Cardiomegaly, Increased Pulmonary Vasculature, Pulmonary Hypertension, and Borderline to Slightly Decreased Systemic Arterial Oxygen Saturation
had but 2 cusps, but in each of these cases one of the cusps was an obvious fusion of 2 cusps. Clinical findings appeared to be unrelated to the type of origin of the pulmonary arteries, whether arising as separate vessels or as a common vessel. None of our cases had pulmonary arteries arising from opposite sides of the truncus. Instead, the vessels arose from the left posterior aspect of the truncus. Case 13 was an exception in this respect, the pulmonary arteries arising from the right posterior aspect of the truncus, but there was also dextrocardia and a left aortic arch in this case. Although the heart was not a true mirror image dextrocardia, it seems plausible to implicate inversion of the truncus structures.

Five of the 14 cases had associated noncardiac defects, but all of the latter were different.

**Summary**

Fourteen cases of true truncus arteriosus of type 1 (with 2 pulmonary arteries) are presented, with emphasis on the variation in clinical findings. Three lived to be 7 years or more of age. Growth retardation, respiratory infections, and dyspnea were common features. Cyanosis was not present in all cases. There was usually a precordial bulge, even in infancy. A systolic thrill and murmur along the left sternal border were typical, with occasionally a blowing diastolic murmur along the upper left sternal border in the older patients, but never a machinery murmur. No instances of a “split” pulmonic second sound were reported. Blood pressures were generally normal. The electrocardiogram usually showed right axis deviation and right ventricular preponderance, but the most consistent finding was peaking of the P waves. There was typically cardiac enlargement with an increase in the pulmonary vascular markings. The pulmonary artery segment was usually flat, but in some cases a “bulge” was formed by a left pulmonary artery, generally with a high take-off. A “shelf” was noted only once in the left anterior oblique view.

A “pure” and accentuated pulmonic second sound appears to be a cardinal diagnostic point. Angiocardiography, preferably biplane, is the most informative of the diagnostic procedures, though in cases where differential streaming results in a near normal systemic arterial oxygen saturation, this test, as well as conventional retrograde aortography, will fail to outline the defect. It is with cases in this group that the clinician is likely to make an erroneous diagnosis. Aortography, with injection of the contrast material through a catheter into the “ascending aorta” should be very informative under such circumstances. Dye-dilution techniques combined with cardiac catheterization should also be very helpful.

The possibility of truncus arteriosus should be considered in all cases resembling “atypical” patent ductus arteriosus, aortic-pulmonary defect, and ventricular septal defects with marked pulmonary hypertension. A slight decrease in systemic arterial oxygen saturation and inability to enter the pulmonary arteries, together with a ready entry into the ascending aorta, should increase this suspicion. The finding of a large and actively pulsating “aorta” at fluoroscopy in a patient with the other cardiac catheterization findings of a ventricular septal defect and right ventricular hypertension is quite suggestive of a truncus arteriosus, since the aorta is invariably relatively small and inconspicuous in a patient with a left-to-right shunt due to isolated ventricular septal defect. Moreover, in a patient suspected of having a patent ductus arteriosus, especially if it is considered somewhat atypical and cardiac catheterization discloses right ventricular hypertension together with a left-to-right shunt at that level, the possibility of a truncus arteriosus must be considered.

**Summario in Interlingua**

Es presentate 14 casos de ver trunco arteriose, typo 1 (con 2 arterias pulmonar). In le presentation, le variabilitate del constatazioni clinic es sublineate. Tres del pacientes viveva a etates de 7 annos o plus. Retardo de crescentia, infecciones respiratorii, e dyspneea esseva aspectos commun. Cyanose non esseva presente in omne casos. In le majoritate del casos un protrusion precordial esseva notate, mesmo in le infantia. Un fremito o murmure systolic al longo del margine sinistro-sterinal esseva typic, con a vices un sufflante murmure diastolic al longo del margine supero-sterinal.
in le patientes de plus alte etates. Nulle rumor a machinas esseva notate. Nulle caso de “bifide” secunde sono pulmonic esseva reportate. Le pressiones sanguine esseva generalmente normal. Le electrocardiogramma exhibiva usualmente deviation dextorose del axe e preponderantia dextero-ventricular, sed le plus uniforme constatation esseva le appuntation del undas P. Allargamento cardiac esseva typic, con augmento del marcas pulmono-vascular. Le segmento del arteria pulmonar esseva usualmente platte, sed in certe casus un protrusione esseva formate per un arteria sinistro-pulmonar, generalmente a initio alte. Un “banca” esseva notate un sol vice, in le vista oblique sinistro-anterior.

Un “pur” e accentuate secunde sono pulmonic es apparenemente un puncto diagnostic cardinal. Angiocardigraphia — preferibilemente blipan—es le plus informante manovra diagnostic, sed in casos in que fluxo differential resulta in quasi normal saturation oxygenic del arterias systemic, iste technica—si ben como le aortographia retrograde conventional —non pote succeder a definir le defecito. Il es in casos de iste gruppo que le clinico curre le plus grande risco de un diagnose erronee. Aortographia—con injection del substantia de contrasto via un catheter a in le “aorta ascendente”—pote devenir multo informative sub tal conditiones. Le mesmo vale pro technicas a dilution de colorantes in combination con catheterisation cardiac.

Le possibilitate de trunco arteriose deberea esser prendite in consideration in omne casos que resimila “atypic” patente ducto arteriose, defecito aorto-pulmonic, e defectos ventriculo-septal con marcate grades de hypertension pulmonary. Leve augmentos del saturation oxygenic in le sanguine arterial systemic e le impossibilitate de entrar in le arterias pulmonar combine con facile accessibilitate del “aorta ascendente” pote servir a reinforziar ille suspicion. Le constatation fluoroscopie de un grande “aorta” a pulsation active in un patienti qui exhibi le altere constatationes de catheterisation cardiac caracteristic de un defecito ventriculare e de hypertension dextero-ventricular es un forte indication de trunco arteriose, proque le aorta es semper relative-mente parve e inconspicue in patientes con derivation sinistro-dextere in consequentia de un isolate defecito ventriculo-septal. In plus, in un patiente in qui il existe le suspicione de patente ducto arteriose—specialmente si illo pare esser un paucu atypic e si le catheterisation cardiac revela hypertension dextero-ventricular insimul con un derivation sinistro-dextere a ille nivello—le possibilitate del presentia de trunco arteriose debe esser prendite in consideration.

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Although cardiac infarction has rarely been observed to occur in patients with active thyrotoxicosis, the authors present 3 such patients discovered during the past 4 years. The clinical course of the 3 patients did not reveal definitive differences between them and others who suffered cardiac infarction in the absence of thyrotoxicosis. The onset of infarction seemed to have been associated with a lesser severity of pain than usual in each of these 3 patients. With the advent of more effective medical measures for suppressing thyroid toxicity, it is conceivable that patients with coronary atherosclerosis and thyrotoxicosis may be relieved of their attacks of angina more readily, but may suffer cardiac infarction when thyroid toxicity is brought under control, unless steps are taken to avoid elevation of serum lipid levels. A further explanation for the rare coincidence of these 2 disorders is to be found in the work recently reported by Rowe et al. They found that the hypermetabolic state of thyrotoxicosis includes the myocardium. When patients were restored to the euthyroid state with treatment, the cardiac output, cardiac work, coronary blood flow, and myocardial oxygen consumption became normal, and the coronary vascular resistance increased.

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Truncus Arteriosus: Clinical Study of Fourteen Cases
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Circulation. 1957;16:586-598
doi: 10.1161/01.CIR.16.4.586
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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