Truncus arteriosus
An anatomical-angiographic study

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SUMMARY A series of 25 cases of truncus arteriosus communis examined post mortem was studied retrospectively. Fifteen of them had had angiographic studies, enabling anatomo-radiographic correlative analysis to be made. All cases had situs solitus of the atria and concordant connections between atria and ventricles. The three main components of the malformation, present in all 25 cases, were: (a) ventricular septal defect, (b) single semilunar valve, (c) anomalies of the aortopulmonary septum.

The ventricular septal defect was always subarterial but its size and its relation to mitral and tricuspid valves were extremely variable. The truncal valve varied greatly also in the number of its cusps and its relation to right and left ventricles. Above valvular level the presence or absence of residual aortopulmonary septum was reflected by the presence or absence of a main pulmonary artery. Additional supratruncal malformations produced variations of the anatomy of the aorta and pulmonary arteries. The angiographic demonstration of all these components was obtained best by selective angiography using special projections.

From this study it appears that there is a wide degree of variability in all of the three main components of truncus arteriosus. Such anatomical variations should be identified angiographically in each patient in order to provide enough information before corrective surgery, but they do not alter the basically homogeneous anatomy of the category of truncus arteriosus and therefore do not justify complex classifications.

Truncus arteriosus communis, a relatively uncommon heart malformation, has appeared to be a sharply defined entity since its original description.1 The malformation is, however, variable in its components if indeed there is more than one component to it. The embryological basis of the defect is not fully understood. Though most classifications try rigorously to follow anatomical variables2−6 the concept of what constitutes essentially the “truncus” from an embryological and anatomical point of view reflects itself in the acceptance or not of specific cases in the category of “common truncus arteriosus”. Truncus arteriosus, however, is now routinely surgically corrected7−9 and one should approach the individual case not on an embryological basis but by carefully studying its various anatomical components with the primary idea of giving the surgeon the information necessary for its repair. These anatomical details can be identified before operation with high accuracy through a careful angiographic examination.

With these ideas in mind, we have reviewed a series of specimens from the anatomical collection of the Department of Pathology at the University of Alabama in Birmingham and compared them with the corresponding postmortem angiographic studies in order to make a retrospective correlative analysis.

Definition of terms

Truncus arteriosus communis is defined in this paper as a congenital heart malformation involving the ventriculoarterial connection in which a single outlet is present and is characterised by: (a) the presence of a single semilunar valve annulus as the only exit from the heart, (b) a subarterial ventricular septal defect, and (c) absence or severe deficiency of the aortopulmonary septum.

There are two related malformations which though close to truncus arteriosus are separate entities; they

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are: (a) the aortopulmonary window, a malformation in which there are two well defined semilunar valves associated with a defect in the aortopulmonary septum, and (b) subarterial ventricular septal defect, a defect located immediately beneath two distinct semilunar valves, the result of absence of infundibular septum.

For specific anatomical structures such as infundibular septum, ventriculoinfundibular fold, trabecula septomarginalis, and aortopulmonary septum, we have followed the definitions given by Crupi et al., recognising that some of them are still far from accepted generally.

Subjects and methods

We examined 25 cases which fit the definition of truncus arteriosus from the anatomical collection at the University of Alabama in Birmingham. The specimens belonged to 17 female and eight male patients. The ages were between 2 days and 71/2 years.

The anatomical study was centred on the analysis of the basic components of the malformation: (a) the anatomy of the truncal valve, (b) the infundibular septal defect, (c) the anatomy of the aortopulmonary septal defect, and (d) any other associated malformation, either of the heart itself or of the great vessels, specifically the aortic arches. The hearts were examined using the sequential approach described by others. Fifteen of the cases in the series belonged to patients who had had cineangiograms performed before their death. The cineangiograms were taken on a 35 mm cine camera at 60 frames per second. Selective opacification of the truncus and right and left ventricles was obtained. Frontal and lateral views were used for truncal angiography and long axial and elongated right anterior oblique views for the ventriculograms. The angiographic anatomy was analysed and compared with the anatomical findings.

Results

ATRIAL MORPHOLOGY

Situs solitus of the atria was present in all cases. Atrial septal defect of the ostium secundum type (patent foramen ovale) was seen in 20 hearts. One of them had an associated common atrioventricular canal defect.

VENOATRIAL CONNECTIONS

The systemic veins were connected normally to the right atrium in 22 cases. In one case there was interruption of the inferior vena cava with aygos continuation. A persistent left superior vena cava, connected to the right atrium through the coronary sinus, was present in two cases. The pulmonary venous connection was to the left atrium in the usual manner in 23 specimens. Two cases had partial anomalous venous return to the right atrium. There was no case of totally anomalous pulmonary venous return.

ATRIOVENTRICULAR CONNECTIONS

Concordant connections between the atria and ventricles were present in all cases. The atrioventricular connection was in parallel fashion, on the same spatial side. The mitral and tricuspid valves were well formed in all cases, except in the one with common atrioventricular canal and in one which showed a pronounced degree of tricuspid hypoplasia and a small hypoplastic right ventricle.

VENTRICULAR SEPTAL DEFECT

All the cases had a ventricular septal defect, immediately beneath the truncal valve. Therefore, the roof of the defect was represented in all cases by the truncal valve itself. The anterior and inferior aspects of the defect were always smooth and muscular, representing the two limbs of the trabecula septomarginalis and the top of the trabecular septum (Fig. 1). The posterior aspect of the defect was variable depending primarily on the size and direction of the posterior arm of the trabecula septomarginalis and its continuity or not with the ventriculoinfundibular fold (Fig. 2). In 20 specimens, there was discontinuity between the ventricular septal defect and the tricuspid annulus as the result of the interposition of these muscular masses. Only in two cases was there a definite continuity between the defect and the tricuspid valve (perimembranous ventricular septal defect). In three cases there was no clear definition of this point.

As seen from the left ventricle, the defect was in front of the mitral valve being separated from it by the ventriculoinfundibular fold and trabecula septomarginalis in 22 hearts (Fig. 2). The mitral valve formed part of the posterior aspect of the defect in three specimens.

The upper rim of the trabecular septum was extrapolated toward the truncal valve in order to analyse the relative relation of the truncal vessel to the two ventricles. The truncal valve was related to the right and left ventricles in identical proportions in 15 specimens (Fig. 3). In five cases more than 50% originated from the right ventricle (Fig. 4 and 5) and in the other five, more than 50% of the circumference was placed to the left (Fig. 2). This relative preference for either ventricle changed from a slight tendency towards one side to practically unique origin from one ventricle in others. The size of the ventricular septal defect was variable but tended to be large (22 specimens), roughly the diameter of the truncal valve itself. In three cases, prominence of one of the elements of the inferior border reduced the size of the
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Fig. 1 The heart has been opened from the right ventricle (RV) into the truncus (T). The small, posterior aorta (AO) ended in two carotid arteries and there was interruption of the aortic arch. Two large pulmonary arteries (RPA and LPA) are seen. A large ductus (D) gave origin to the descending aorta (DA) which produced the two subclavian arteries. Notice the inferior borders of the ventricular septal defect (VSD) bound by the arms of the trabecula septomarginalis (TSM) with absence of the infundibular septum. The truncal valve (TV) is almost normal. The remaining aortopulmonary septum is represented by the line of asterisks.

defect and displaced it anteriorly or posteriorly (Fig. 4 and 5).

The ventricular septal defect was shown on right and left ventriculograms on a long axis view and on the right ventriculogram in the elongated right anterior oblique view. In the elongated right anterior oblique view (Fig. 6) it is seen during systole as a thick column of contrast medium on the superior and left border of the right ventricular chamber. The defect itself is bound by the junction of the ventriculoinfundibular fold with the posterior arm of the trabecula septomarginalis posteriorly, by the truncal valve superiorly, and by the free border of the superior arm of the trabecula septomarginalis on the left. The left contours are bound in some patients by septal trabeculations rather than by the trabecula septomarginalis itself. In cases where the ventricular septal defect was in continuity with the tricuspid valve annulus, the posterior arm of the trabecula septomarginalis was small or absent.

In the right ventriculogram in the long axis view (Fig. 7) the right ventricle is seen emptying into the truncal channel. This connection is bound posteriorly by the septal structures and anteriorly by the ventriculoinfundibular fold.

The long axis view of the left ventriculogram (Fig. 8) delineates the ventricular septal defect, sited immediately beneath the truncal valve in the area where the infundibular septum should be placed. The left ventricular outflow tract is seen formed by the anterior mitral leaflet posteriorly and by the trabecular septum anteriorly. This view also shows the mitral-truncal valve continuity when present. The right ventricle is usually opacified partially on the left ventriculogram through the ventricular septal defect. In some cases this fact allows us to study the relation.
between the mitral and tricuspid valves with the truncal valve. This view is also of particular help in cases when the origin of the truncus is primarily from the left ventricle.

TRUNCAL VALVE

The annulus was moderately enlarged in all cases, except two. The truncal valve had four cusps in 10 hearts, three in 11, two in one, and was unspecified or poorly defined in three. The free borders of the aortic cusp were thick and irregular (dysplastic) and the whole valvular apparatus deformed in 15 hearts (Fig. 4 and 5). Eight cases had normal appearance of the cusps (Fig. 1 and 3). In two, the valve had been removed. The semilunar valve was in continuity with the mitral valve in 20 specimens (Fig. 2 and 3) and it was separated by muscular structures in five. The truncal valve was in discontinuity with the tricuspid valve in 21 hearts and was part of the annular fibrous tissue in four. Regurgitation and stenosis of the semilunar valve were considered to be difficult to assess in the anatomical specimens and no attempt to give specific figures about this point were therefore undertaken.

The angiographic assessment of the valve is made through a selective truncal angiogram (Fig. 9) though it is frequently well visualised on right and left ventricular angiograms as shown in Fig. 7 and 8. The well formed tricuspid truncal valve can be identified using frontal and lateral views. The cusps are better seen in diastole as round smooth contours forming the base of the large single vessel. When the valve is bicuspid, the appearance is that of a two valved structure. Dysplastic leaflets are seen as irregular borders with poorly defined cusps (Fig. 10). Their borders are usually thickened and can be identified easily in motion pictures. Many of these dysplastic valves will be quadricuspid, but it is only occasionally that a definite
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Fig. 3 A similar photograph of another specimen. The ventricular septal defect (VSD) leaves about 50% of the truncal valve (TV) to each ventricle. The valve is tricuspid and practically normal. It has continuity with mitral (MV) and tricuspid (TV) elements. The truncus (T) divides immediately into two well recognisable vessels. A large opening to the left and anteriorly represents the pulmonary artery (P) which had been closed by a Teflon patch at surgery (not removed) and connected to the right ventricle by an external conduit. The open vessel to the right represents the aorta (AO). The sizes of the aorta and pulmonary artery are similar. LV, left ventricle, RV, right ventricle.

Fig. 4 Heart and truncus opened from the right. The truncus (T) produces a predominant aorta (AO) and two independent small pulmonary arteries (RPA) (LPA), implying absence of the aortopulmonary septum. They originate shortly above a much deformed, dysplastic, and quadricuspid truncal valve (TV). The origin of the truncus is almost exclusively from the right ventricle (RV). The defect (VSD) is fairly small because of the hypertrophy of the components of the trabecula septomarginalis.

Fig. 5 Right ventricular view of an example of truncus type I with similar size of the aortic (AO) and pulmonary (P) components. The truncal valve (TV) is tricuspid but badly deformed and redundant. It lies mostly to the right of a very small ventricular septal defect (VSD). The small ventricular septal defect results from hypertrophy of the superior arm of the trabecula septomarginalis (TSM). There is no infundibular septum. The remaining aortopulmonary septum is indicated by the asterisks. RV, right ventricle.
Fig. 6 Systolic phase of right ventriculogram in elongated right anterior oblique view. The right ventricle (RV) empties into the truncus (T) through a large ventricular septal defect (arrowheads). Notice that the defect is separated from the tricuspid valve annulus (tv) by a large filling defect (arrow) which represents the ventriculo-infundibular fold. The aorta (AO) and the pulmonary artery (P) are of similar size in this example. TSM, trabecula septomarginalis.

Fig. 7 Right ventriculogram in long axial view. (A) Systole and (B) diastole. The right ventricle (RV) is oriented inferiorly and to the right. The tricuspid valve (tv) is separated from the truncal valve (clear white arrows) by the ventriculo-infundibular fold (top black arrow). The posterior contour of the ventricle is formed by the trabecular septum inferiorly and by the trabecula septomarginalis superiorly (low black arrow). The truncal valve is well seen on diastole, but it has restricted opening on systole. The aortopulmonary septum (large white arrow) is seen separating the aorta (AO) from the pulmonary artery (P). T, truncus.
Fig. 8  Left ventriculogram in long axial view. (A) Systole and (B) diastole. The truncal valve (arrows) is supported by both ventricles in approximately equal proportions. The truncus arteriosus (T) is seen immediately above the valve. The subarterial ventricular septal defect (vsd) is just beneath the truncal valve. RV, right ventricle; LV, left ventricle; TS, trabecular septum; mv, mitral valve.

Fig. 9  Truncal angiogram in frontal view. A case with widely separated origin of the pulmonary arteries. The truncal valve (white arrows) is tricuspid and well formed. The left pulmonary artery (LPA) originates earlier (black arrows) and more frontally than the right (RPA and arrowheads) producing a negative image which does not represent true aortopulmonary septum. The aorta (AO) shows a right sided arch. T, truncus.
assessment of the number of cusps may be obtained.

Stenosis or regurgitation of the valve is evaluated in a similar way to that used in normal ventriculoarterial valves. A reflux through the valve in diastole towards one or both ventricles is seen in regurgitation, while a small orifice, out of proportion to the diameter of the vessel just above the valve, is seen in stenosis (Fig. 10).

CORONARY ARTERIES
The anatomy of the coronary arteries was extremely variable. It depended a great deal on the number of truncal cusps and the orientations of the valve. On the other hand, no major anomaly of these vessels was identified in this series.

AORTOPULMONARY SEPTUM
The aortopulmonary septum was absent in the area immediately above the truncal valve in all cases. Twelve cases showed absence of only a very small portion of the septum and a definite main pulmonary artery could be seen originating from the truncus (type I of Edwards) (Fig. 3 and 5). The predominant vessel above the area of aortopulmonary septal absence was an aorta in 10 cases. In two cases the main vessel represented a pulmonary artery, and there was a small aortic trunk originating to the right and posterior to the truncal vessel (Fig. 1). The origin of the main pulmonary artery in the cases with dominant aorta was always anterior and left, immediately above the truncal cusps. The artery coursed in a normal way, upwards, superiorly, and posteriorly until it bifurcated. The left pulmonary artery was always higher than normal. In two cases, the left pulmonary artery had an independent origin as a continuation of a ductus arteriosus and, therefore, only one pulmonary artery, the right, originated from the truncus as a continuation of the main pulmonary artery (Fig. 11).

Much more extensive absence of the aortopulmonary septum reflecting as an independent origin for

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Fig. 10  Left ventriculogram in long axial view in a case of truncus with associated aortic arch interruption. (A) Systole and (B) diastole. The aortic arch interruption (arrow) results in conspicuous enlargement of the pulmonary artery (PA) as compared with the aorta (AO). The linear negative image between the two is the aortopulmonary space. Below its lowest point the aortopulmonary septum is missing. The descending aorta (DA) originates from the present ductus arteriosus (D). A restricted ventricular septal defect (white arrow) is well seen in diastole. The dysplastic truncal valve shows as filling defects (arrowheads) in the valvular area as the valve opens in systole. There is also suggestion of valvular stenosis. LV, left ventricle; T, truncus.
this structure varies depending on the ratio in size between the aorta and the pulmonary artery. Absence of this line is seen when the right and left pulmonary arteries arise independently and directly from the truncus, albeit a negative shadow separating right and left pulmonary arteries may erroneously suggest aortopulmonary septum (Fig. 9).

DISTAL VASCULAR STRUCTURES
The relative size of the aorta and pulmonary artery above the aortopulmonary septal defect varied considerably, though the aorta was almost always larger. Only in four cases did the main pulmonary artery approximate the size of the aorta (Fig. 5). The aorta was sited to the right of the main pulmonary artery in all cases. The aortic arch was right sided in eight specimens (Fig. 11) and left sided in 17.

The ascending aorta was much smaller than the pulmonary artery in two specimens. These two cases had an associated interruption of the aortic arch (Fig. 1) which was probably the cause of the selective hypertrophy of the pulmonary component of the great vessels. Moderate degrees of hypoplasia of the distal aortic arch were found in 10 cases, but no real coarctation was identified. A persistent ductus arteriosus was present in eight cases, including the two with predominance of the pulmonary artery where the ductus was larger than the ascending aorta (Fig. 1). In two cases, the origin for the left pulmonary artery was the ductus arteriosus with discontinuity of the pulmonary arteries (Fig. 11). Anomalous origin of the right and/or left subclavian arteries was seen in four cases. In one of the cases with predominance of the pulmonary artery, both subclavians originated from the distal part of the aorta, while in the other, the right subclavian originated from the same place taking a retrooesophageal course.

Peripheral pulmonary stenosis was seen in two cases; one on the right and one on the left, just a few centimetres above the origin from the truncal vessel. The distal intraparenchymatous pulmonary arterial anatomy appeared normal by angiographic and anatomical observation.

Truncal angiography is the method of choice to show the variations of the great vessels above the truncal defect proper (Fig. 9); these changes, however, are frequently well identified on right or left ventriculograms (Fig. 7 and 10).

Discussion
Truncus arteriosus communis was first described by Buchanan1 more than a century ago.

As knowledge about the embryological development of the bulbo-truncal area increased, the definition of what constituted “truncus arteriosus”
changed accordingly which reflected in the different classifications proposed for this malformation.\textsuperscript{2-6} Since 1970, truncus arteriosus has been successfully surgically repaired.\textsuperscript{7} It is evident that a proper preoperative definition of the anatomical details will help in the surgical approach. For that purpose, it is important to define the condition on an anatomical basis.

Minor variations are present in different definitions\textsuperscript{5-6} but we feel that of Crupi et al.\textsuperscript{10} is anatomically accurate: "A single arterial trunk that leaves the heart by way of a single arterial valve and that gives rise directly to the coronary, systemic and one or both pulmonary arteries." We also feel that a much briefer way to define this condition is that of: "a defect in the separation of the ventricular outlets of the heart involving supravalvular, valvular, and infravalvular levels". From this definition, three distinct components of the malformation are always present: (a) a single truncal valve, (b) a subarterial ventricular septal defect\textsuperscript{14} in the presence, most frequently, of two definite, identifiable right and left ventricles, and (c) a deficiency in the aortopulmonary septum which may be more or less extensive in such a way that a definite recognisable main pulmonary artery may or may not be present.

(a) The presence of a single arterial valve is an important component in truncus arteriosus; its morphology has been extensively described.\textsuperscript{5-6,15} By using the term "single", it should be understood that we do not imply the exclusion of the concept of "common" valve. Instead we feel that the truncal valve is both single and common. The truncal valve is usually related to the right and left ventricle in equal proportion but some may be mostly related to the right or left ventricles as described in this series. This fact has surgical implications related to the construction of the new left ventricular outflow tract.

(b) The infundibular ventricular septal defect is in a subarterial position, roofed by the truncal valve instead of the aortic and pulmonary valves.\textsuperscript{14} We support the view that the defect is the result of the absence of infundibular septum,\textsuperscript{6,10} leaving exposed the trabecula septomarginalis and the ventriculoinfundibular fold as the boundaries of the defect. The size of the defect varies but in our opinion it is always present. The report by Carr et al.\textsuperscript{16} who described a case of truncus arteriosus without ventricular septal defect is similar to the cases that Van Praagh selects for the separation of his type B truncus arteriosus without ventricular septal defect.\textsuperscript{3-5} In our opinion, such cases are either aortopulmonary windows, or cases of true truncus arteriosus in which a low implantation of the truncal valve prevents the interventricular communication even though the defect exists as a result of the absence of infundibular septum.

Truncus arteriosus occurs usually in hearts with two well recognisable left and right ventricles. Our series consists exclusively of such cases, though one of them had relative hypoplasia of the right ventricle. The condition "truncus" could, however, occur in cases where one of the chambers may be considered only as an accessory chamber or even from a single ventricle with no ventricular partition whatsoever.

(c) The aortopulmonary septum is always deficient in truncus arteriosus immediately above the truncal valve. When a remnant aortopulmonary septum is present, the main pulmonary artery is recognised and defines one variety of truncus arteriosus (type I). If the aortopulmonary septum is completely absent, the right and left pulmonary arteries arise from the truncus itself and this fact defines the second variety of truncus arteriosus (type II).

The majority of cases will have an aorta larger than the pulmonary artery. There will be cases, much less common, where the pulmonary artery will be larger than the aorta. This phenomenon will occur when there are associated anomalies of the aortic arch requiring the presence of a large ductus and the overgrowth of the pulmonary artery. We do not subdivide the types of truncus arteriosus by their associated lesions as others have suggested.\textsuperscript{3-5} We believe that "truncus arteriosus" should be defined in its own terms and according to very strict guidelines. Any added anomaly that changes the pattern of blood flow will change the anatomical appearance of the heart, but will not be essential in the malformation we call "truncus arteriosus".

This study has shown that axial angiography is able to show the anatomical details of the truncal anomaly itself in all its basic components as well as the associated malformations commonly present, providing accurate information about all points necessary for surgical follow-up in the individual case.

References

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