Anatomical study of truncus arteriosus communis with embryological and surgical considerations

Gaetano Thiene, Uberto Bortolotti, Vincenzo Gallucci, Vito Terribile, and Piero A. Pellegrino

From the Departments of Pathology, Cardiovascular Surgery, and Paediatrics, Medical School, University of Padua, Padua, Italy

Twelve specimens of truncus arteriosus communis have been studied anatomically, with special reference to the conal anatomy and to the associated cardiac anomalies which can create additional problems if surgical repair is planned. A wide spectrum of conal morphology has been observed, suggesting that differential conal absorption is a developmental characteristic of truncus arteriosus as well as of transposition complexes. The invariable absence of septation of the ventricular infundibula and semilunar valves, in spite of the variable anatomy of the free wall of the conus, indicates that all types of truncus arteriosus, ontogenetically, should be considered as a single undivided conotruncus. Various types of ventricular septal defect were found: (a) ventricular septal defect with absent crista, in which no remnants of conal septum are present; (b) supracristal ventricular septal defect, in which vestigial conal septum is seen in front of the membranous septum; (c) bulboventricular foramen, associated with univentricular origin of the truncus from the right ventricle. Frequent associated anomalies are underdevelopment of the aortic arch, truncal valve malformations, and obstructive ventricular septal defect. The AV conduction system studied in one case showed an arrangement similar to Fallot's tetralogy with the His bundle and the left bundle-branch in a safe position behind the posteroinferior rim of the defect. The postoperative fate of the frequently abnormal truncal valve and the theoretical indications for total repair for Type IV truncus are also discussed.

The anatomical study of truncus arteriosus communis has gained great importance since total surgical correction of this malformation has become possible (McGoon, Rastelli, and Ongley, 1968; Mair *et al.*, 1974). Recent reports have emphasized that the problems facing the surgeon are related not only to the lack of partitioning between aorta and pulmonary artery, but also to the status of the truncal valve, the degree of aortic arch development, and the size of the ventricular septal defect (Becker, Becker, and Edwards, 1971; Gomes and McGoon, 1971; Mair *et al.*, 1974; Bharati *et al.*, 1974).

In addition, the morphological study of a lesion such as truncus arteriosus communis, in which partial or complete absence of division of the conotruncus is present, may help to clarify which developmental structures take part in the septation of the outflow tracts of the normal heart. With this in mind, we have also examined cases presenting with aortopulmonary septal defect or conal ven-tricular septal defect which are developmentally related to truncus arteriosus.

Definition of terms

The term *conus*, embryogenetically, refers to the middle third of the bulbus cordis; it is formed by free walls (parietal conus) and by septum (conal septum) derived from the fusion of the conus ridges. Anatomically, the conus corresponds to the outflow tracts of both ventricles and conventionally it refers to the muscular cardiac segments interposed between the semilunar and atrioventricular valves; the conal septum is that portion of the ventricular septum which separates the ventricular infundibula. In the early stages of cardiac development, the atrioventricular channel is separated from the semilunar valves by the parietal conus; a process of differential absorption leads to direct fibrous continuity between mitral and aortic valves, while

conal muscular discontinuity persists between pulmonary and tricuspid valves.

The term *truncus* indicates the distal bulbus cordis, the site of growth of the truncal ridges. Anatomically, the truncus corresponds to the semilunar valves; the truncal septum, embryogenetically derived from the fusion of the main truncal ridges, corresponds anatomically to the fibrous septum which separates the roots of the aorta and pulmonary artery. The ascending aorta and main pulmonary trunk originate from the ventral aorta (or aortic sac) and are septated, during development, by the aortopulmonary septum. The limits of truncus and aortic sac and of truncal and aortopulmonary septa are unknown. *Conotruncus* is a collective term used for the conus and the truncus, as is truncoconal septum for the conal and truncal septa.

The *crista supraventricularis* is a muscular segment of the right ventricle interposed between the tricuspid and pulmonary valves: it is a conal structure, consisting of the conal septum medially and of a parietal band derived from the right portion of the conoventricular flange, laterally. The conoventricular flange is the inner curvature of the primitive heart tube, after d-looping, and represents part of the posterior wall of the primitive undivided conus; it is thus an anatomical landmark of the parietal conus. The *septomarginal band* is a muscular trabecula of the right ventricle, formed by the septal and moderator bands, quite distinct from the crista supraventricularis, because from a developmental point of view it is not a conal structure.

Subjects and methods

Of the 400 anatomical specimens of congenital heart diseases collected during the past 10 years in the Institute of Pathologic Anatomy of Padua University Medical School, 12 cases (3%) presented with the classical features of truncus arteriosus communis, i.e. only one arterial trunk, arising from the heart above a ventricular septal defect and giving rise to the coronary, systemic, and pulmonary circulations. Cases with pseudotruncus aorticus and pulmonalis in which the pulmonary trunk and the ascending aorta, respectively, were hypoplastic, as a result of atresia of the correspondent semilunar valve, were excluded. In addition, hearts with truncus arteriosus were compared with those with aortopulmonary septal defect or with conal ventricular septal defect, in which an isolated defect of septation of the outflow tracts of the heart was present.

The truncus arteriosus specimens were studied with reference to the patterns of origin of the pulmonary arteries; the development of the aortic arch and ductus arteriosus in relation to the type of arterio-ventricular alignment; the status of the truncal valve; the type and size of the ventricular septal defect and the morphology of the ventricular outflow tracts; the conducting tissue; the degrees of failure of septation and the pulmonary vascular bed.

Findings

Patterns of origin of pulmonary arteries

Using the classification of Collett and Edwards (1949), 7 cases presented with the pulmonary arteries arising from a short common pulmonary trunk (Type I). In 3 cases, the pulmonary arteries arose independently, in one case posteriorly, in another anteriorly, and in the last case on the left lateral side of the truncus arteriosus (Type II) (Fig. 1). No cases of Type III were observed, while 2 were Type IV with the pulmonary circulation supplied by arterial branches arising from the descending aorta (Fig. 2).

Development of aortic arch and ductus arteriosus in relation to type of arterio-ventricular alignment

Four cases showed a right aortic arch, and one of these presented with an aberrant retro-oesophageal left subclavian artery. In 9 cases, the truncus arteriosus communis had a biventricular origin. In the other 3 it arose entirely from the right ventricle; in 2 of these a left aortic arch was interrupted between the left carotid and subclavian arteries (Fig. 3a), and in the third a left aortic arch was hypoplastic.

The ductus arteriosus was absent in 8 cases and hypoplastic in 1; in the remaining 3, the ductus was widely patent so that there was anatomical continuity between the heart and the descending aorta (Fig. 3a).

Status of truncal valve

The truncal valve was bicuspid in 2 cases, tricuspid in 8, and quadricuspid in 2. The width and the length of the cusps were always unequal. In 10 cases the cusps had a fairly prominent nodular thickening along the free edge.

In a specimen with univentricular origin of the truncus from the right ventricle, the grossly nodular quadricuspid truncal valve was severely regurgitant, the right ventricular cavity was widely dilated and 'jet lesions' were present on the endocardium
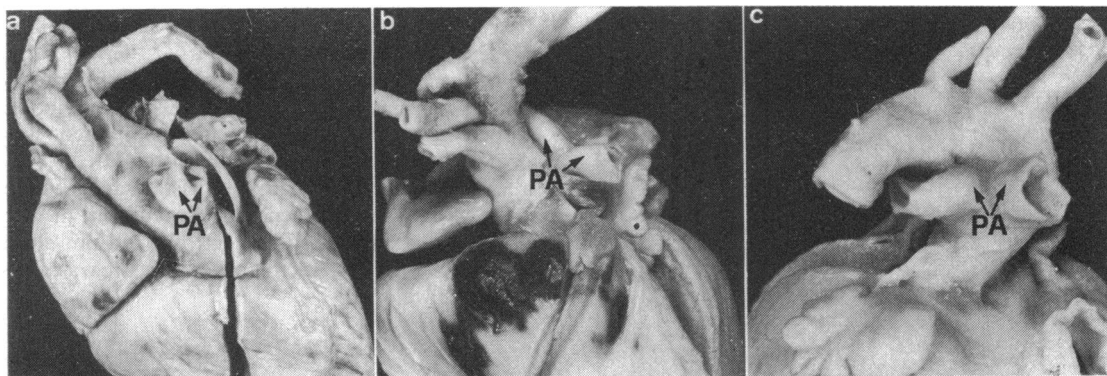
FIG. 1 *Various patterns of origin of pulmonary arteries (PA) in Type II truncus: (a) from anterior aspect of truncus, viewed from the front, (b) from left lateral aspect of truncus, viewed from the front, and (c) from posterior aspect of truncus, viewed from the back.*

(Fig. 3b). Histological examination of this valve showed conspicuous thickening by mucoid connective tissue (Fig. 4).

In a case of Type IV truncus, observed in a 29-year-old man, the tricuspid truncal valve showed moderate fusion of the cusps at the commissures and nodular calcific thickening, causing truncal valve stenosis (Fig. 5).

Type and size of ventricular septal defect in truncus arteriosus communis and morphology of ventricular outflow tracts

Since in all 12 specimens the ventricular septal defect was located anteriorly, corresponding to the conal component of the ventricular septum, it could be regarded as a conal ventricular septal defect (Fig. 6a).

The case material was classified according to the morphology of the margins of the defect, as follows:

(a) Six cases in which the posterior margin consisted of the membranous septum, and the anterior margin consisted of the anterior conal free wall suggesting the term of 'absent crista ventricular septal defect' (Goor *et al.*, 1970) (Fig. 6b). This type is very similar to the classical ventricular septal defect observed in Fallot's tetralogy, with the difference that in this last situation the anterior margin is represented by the deviated conal septum.

(b) Three cases in which conal musculature, located in front of the membranous septum, formed the posterior rim of the defect. In this situation the ventricular septal defect may resemble a supracristal defect (Goor *et al.*, 1970) (Fig. 6c, 10a).

(c) Three cases where the roof of the defect consisted either of a conal vestige of the bulboventricular flange (1 case) (Fig. 6d) or of the anterior mitral leaflet (2 cases) (Fig. 3c) because of the right lateral displacement of the truncus. Here the ventricular septal defect constitutes the only outlet for the left ventricle, as occurs during development before the conotruncal shift towards the midline (Goor, Dische, and Lillehei, 1972; Anderson *et al.*, 1974b). This
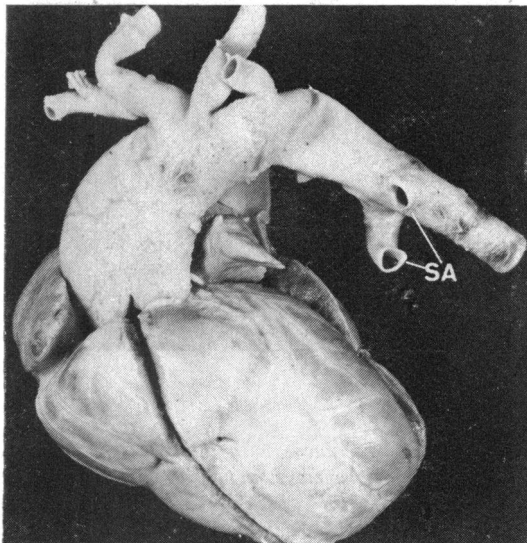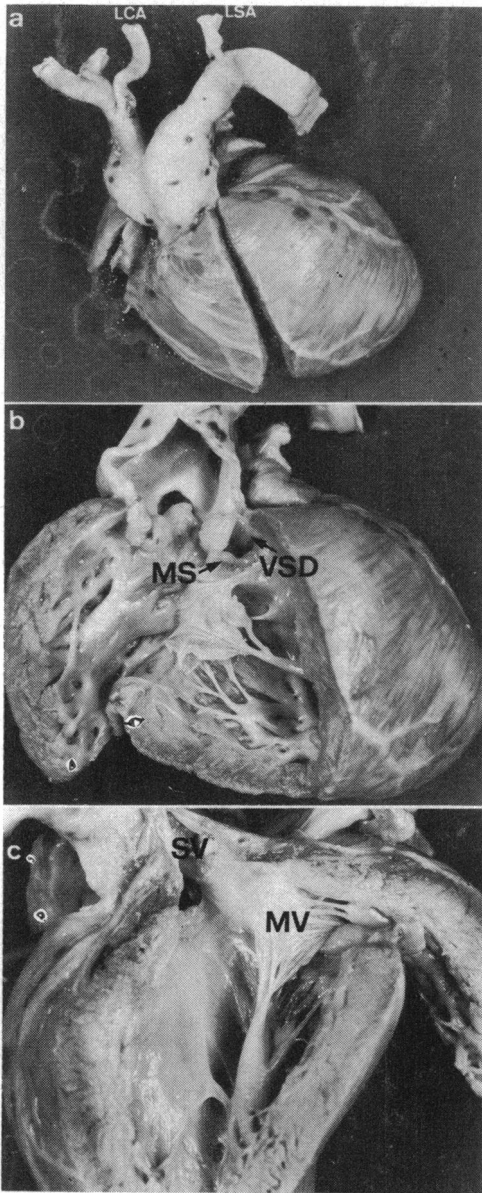

FIG. 2 *Type IV truncus with two large systemic arteries (SA) taking origin from the descending aorta and supplying the right and left lungs.*

FIG. 3 Type I truncus with interrupted left aortic arch. (a) External view showing complete interruption between left carotid (LCA) and left subclavian (LSA) arteries. (b) Right ventricle: truncus arises entirely from right ventricular cavity; quadricuspid truncal valve with thick and nodular leaflets; right ventricle is very dilated, as a result of truncal valve regurgitation, and shows endocardial 'jet lesions';

type of defect may be regarded as a true bulbo-ventricular foramen.

The atrioventricular component of the membranous septum was completely developed in all 12 specimens. The interventricular component was normal in the 3 cases with supracristal defect (Fig. 6a, c) and in one case of absent crista defect, where it appeared as a formed fibrous curtain in the posterior quadrant of the defect, but did not close the defect (Fig. 3b). In the other cases it was only partially developed.

FIG. 4 Histological appearance of truncal cusp, showing abnormal shape and severe mucoid thickening. AW = aortic wall. (Elastic-Van Gieson stain × 12.)

pars interventricularis of the membranous septum (MS) is normally developed, but is 'free floating' and forms the posterior rim of the ventricular septal defect (VSD). (c) Left ventricle: black probe, below semilunar valve (SV), indicates ventricular septal defect which is only outlet for left ventricle; mitral valve (MV) is in fibrous continuity with semilunar valve of truncus and forms roof of defect.

FIG. 5 *Truncal valve from 29-year-old patient with Type IV truncus: there is mild commissural fusion and nodular calcific thickening of the cusps, whose excursion is severely limited, resulting in valvular stenosis.*

Assuming that muscular structures between truncal and atrioventricular valves represent conal free walls, 6 specimens showed fibrous parietal coni, with bilateral direct continuity between the truncal and the atrioventricular valves (Fig. 7a). A more or less developed right parietal conus, with muscular discontinuity between the truncal and tricuspid valves and fibrous continuity between the truncal and mitral valves, was observed in 5 specimens. The right conal muscle often appeared as the 'crista supraventricularis', much as in the normal heart (see Fig. 10). A bilateral parietal conus was present in one case as a muscle cuff (conoventricular flange) separating the truncal valve from both atrioventricular valves (Fig. 7b).

A conal papillary muscle (Lancisi's muscle) was observed in 5 specimens. There was no correlation between its presence and the type of ventricular septal defect.

A pulmonary infundibulum, even blind or potential, was never identified as a separate entity in any of the types of truncus.

In two of the three cases with underdeveloped aortic arch and truncus arising entirely from the right ventricle, there was fibrous continuity between the mitral and truncal valves (Fig. 3c) and in the third there was one muscular parietal conus between mitral and truncal valves (Fig. 7b).

Conducting tissue

In the case with bilateral parietal conus, the conoventricular flange was responsible for a partial obstruction of the ventricular septal defect (Fig. 6d). Serial histological examination of the con-

ducting system showed a normally located compact AV node and the penetrating bundle passing on the left side of the central fibrous body. The branching portion of the AV bundle ran beneath the membranous septum (Fig. 8a) and passed on the left side of the crest of the still intact ventricular septum (Fig. 8b). The left bundle-branch emerged as a continuous sheet before the branching bundle reached the posteroinferior rim of the ventricular septal defect. The right bundle-branch was the direct continuation of the branching bundle (Fig. 8c): it was first observed at the posterior aspect of the ventricular septal defect, on the left of the inferior border of the defect, and penetrated the myocardium to reach the septomarginal band after a long intramyocardial course (Fig. 8d, e, f).

Degrees of failure of septation

The conal component of the ventricular septum was partially or totally absent in all truncus arteriosus communis specimens, indicating lack of septation of the ventricular outflow tracts resulting from conal growth failure. In addition, the semilunar valves and the sinuses of the great arteries were always undivided, suggesting developmental absence also of the truncal septum.

While in Type I truncus the absence of septation involved the ventricular outflow tracts, the semilunar valves, and the sinuses of the great arteries, in the other cases (Types 2 and 4) there was also no division between ascending aorta and pulmonary trunk.

Examination of those specimens showing defects which we consider developmentally related to truncus arteriosus communis disclosed normal septation of the ventricular infundibula, the semilunar valves, and the aortopulmonary sinuses in aortopulmonary window, along with a greater or lesser degree of growth failure in the wall normally separating the ascending aorta from the pulmonary trunk (i.e. aortopulmonary septum) (Fig. 9a). In addition, partial or complete failure of division of the ventricular outflow tracts was observed in the conal ventricular septal defects, though the semilunar valves and the great arteries were normally divided (Fig. 9b).

Pulmonary vascular bed

Patients with type 1 and 2 truncus at death ranged in age from 2 days to 3 months. In each case histological examination of the lungs revealed grade 1 changes of the small arteries and arterioles (Heath and Edwards, 1958).

The age at death of the two patients with Type IV truncus was 1 and 29 years; the former died

with signs of congestive heart failure, and the latter as a result of cerebral abscess and severe hypoxia. The lungs were available for gross and histological examination in the first case only. Gross examination of the heart showed conspicuous dilatation of the left ventricle, suggesting the presence in life of a large left-to-right shunt even though no true pulmonary arteries took origin from the common trunk. The pulmonary circulation was sustained by two large systemic arteries arising from the descending aorta, one for each lung (Fig. 2). The left lung circulation was visualized by post-mortem injection of radio-opaque material into the large systemic artery, and a normal lobar distribution without tortuosity was observed. Histological examination showed no abnormalities in the arterial wall but there was medial hypertrophy of pulmonary arterioles.
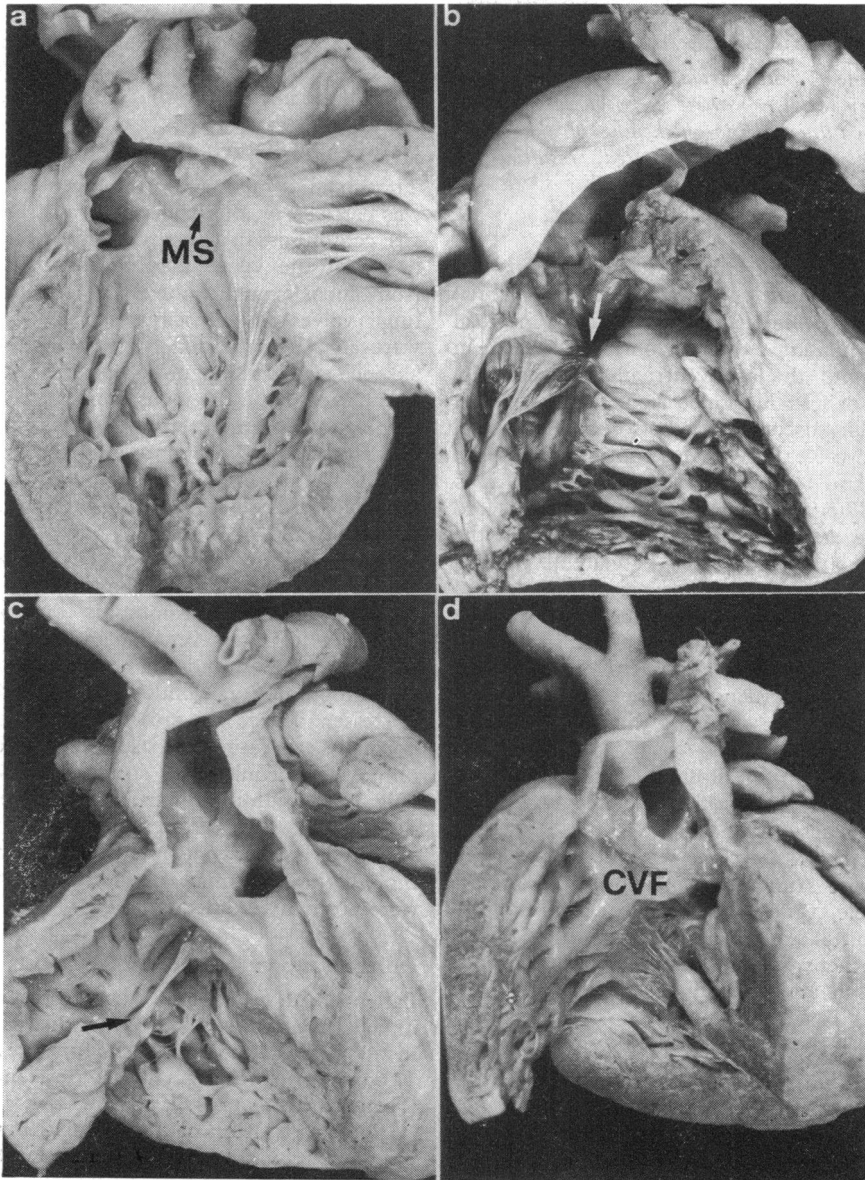
Discussion

Conal anatomy in truncus arteriosus communis and its morphogenetic significance

The morphology of the ventricular outflow tract in truncus arteriosus communis is variable. The posterior wall is usually fibrous with direct continuity between the truncal and the mitral and tricuspid valves, indicating that the embryonic conoventricular flange has undergone complete absorption (Goor et al., 1972; Anderson et al., 1974b). Rarely, this structure is seen as a muscular mass interposed bilaterally between the truncal valve and the AV valves. In the other specimens, while there is fibrous continuity between truncal and mitral valves, a muscular structure separates the truncal from the tricuspid valve, often resembling the crista supraventricularis of the normal heart. Different degrees of infundibular muscular development may also be seen in the anterior wall of the common outflow tract. These findings suggest that a wide spectrum of conal anatomies exists in truncus arteriosus communis and that a differential conal absorption is a developmental characteristic of this malformation as well as of the transposition complexes (Goor and Edwards, 1973; Anderson et al., 1974a; Thiene, Razzolini, and Dalla-Volta, 1976).

Various patterns of truncoventricular alignment may be observed in truncus arteriosus communis. The fact that univentricular origin of the truncus from the right ventricle may be associated with either the persistence of the left portion of the conoventricular flange or its complete disappearance, suggests that the process of differential conal absorption, which normally occurs during the developmental shaping of the infundibula (Goor et al., 1972; Anderson et al., 1974b), is not the only factor playing a role in the transfer of the posterior conus above the left ventricle, and may not be involved at all. Therefore, as we recently suggested in a study on transposition complexes (Thiene et al., 1976), differential conal absorption may not influence the arterioventricular alignment.

Truncus arteriosus communis: extreme hypoplasia or absent septation of conus?

In spite of the variability of conal anatomy, the basic defects in truncus arteriosus communis are always in a high anterior ventricular septal defect and a common semilunar valve, indicating lack of septation of the embryonic conotruncus. For this reason, truncus arteriosus communis may be defined, morphogenetically, as a single undivided conotruncus (M. V. de la Cruz, 1975, personal communication).

The view propounded by Van Praagh and Van Praagh (1965) that truncus is a pseudotruncus aorticus (pulmonary atresia + ventricular septal defect), developmentally determined by an extreme hypoplasia of the subpulmonary conus, is not supported by our finding in one case of a well-formed bilateral muscular conus.

The fact that there are three or four truncal valve cusps more commonly than two has been used in support of the view that the truncal valve might be an aortic valve (Van Praagh and Van Praagh, 1965). We believe that if the main truncal ridges do not develop at all a bicuspid truncal valve results; if one or both main truncal ridges grow without fusing, the resulting cusps are three or four.

What is crista supraventricularis in truncus arteriosus communis?

The muscular structure intervening in five of our cases of truncus arteriosus communis between

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← FIG. 6 *Various types of ventricular septal defect in truncus. (a) Left ventricular view of specimen, showing high anterior location of defect corresponding to conal component of ventricular septum, and in front of membranous septum (MS). (b) Absent crista ventricular septal defect: posterior rim of defect is represented by confluence (white arrow) of the anterior and septal leaflets of the tricuspid valve with membranous septum and medial cusp of mitral valve. (c) Supracristal ventricular septal defect: a muscular structure located in front of the membranous septum forms posterior edge of ventricular septal defect; black arrow indicates hypoplastic anterior right papillary muscle. (d) Bulboventricular foramen: truncus arises entirely from right ventricle and ventricular septal defect is only outlet for left ventricle; because of persistence of conoventricular flange (CVF), roof of defect is formed by conal muscle; resulting ventricular septal defect is partially obstructive and truncal valve is bicuspid. Because of these anatomical features, this specimen is true fetal common conotruncus (absent truncal and conal septation, persistence of conoventricular flange, and univentricular origin of truncus from bulbus cordis).*
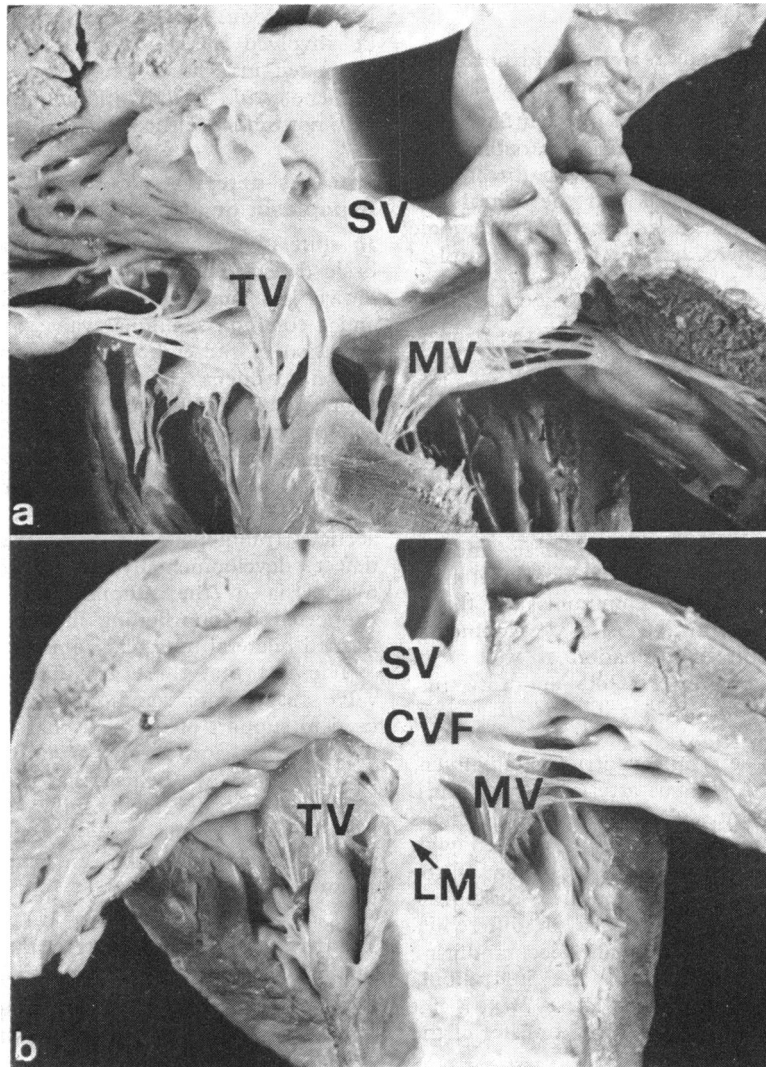
FIG. 7   *Contrasting features of ventricular outflow tracts in truncus arteriosus communis.* *(a) Both atrioventricular valves are in complete fibrous continuity with the truncal valve; posterior edge of ventricular septal defect is represented by interventricular part of membranous septum; no muscle of Lancisi; posterior walls of both ventricular outflow tracts are fibrous, indicating complete absorption of conoventricular flange; no vestige of conal septum exists.* *(b) In contrast to previous case, both ventricular outflow tracts in this specimen are muscular; large cuff of muscle represents persistent conoventricular flange (CVF) and separates truncal valve from both AV valves; posteroinferior edge of ventricular septal defect is formed by confluence of mitral and tricuspid septal leaflets with membranous septum; truncal valve is bicuspid and truncus arises entirely from right ventricle (same specimen as Fig. 6d); 'conal' papillary muscle is seen, though conal septum is totally absent. LM=Lancisi's muscle, MV= mitral valve, SV=semilunar valve of truncus, TV=tricuspid valve.*
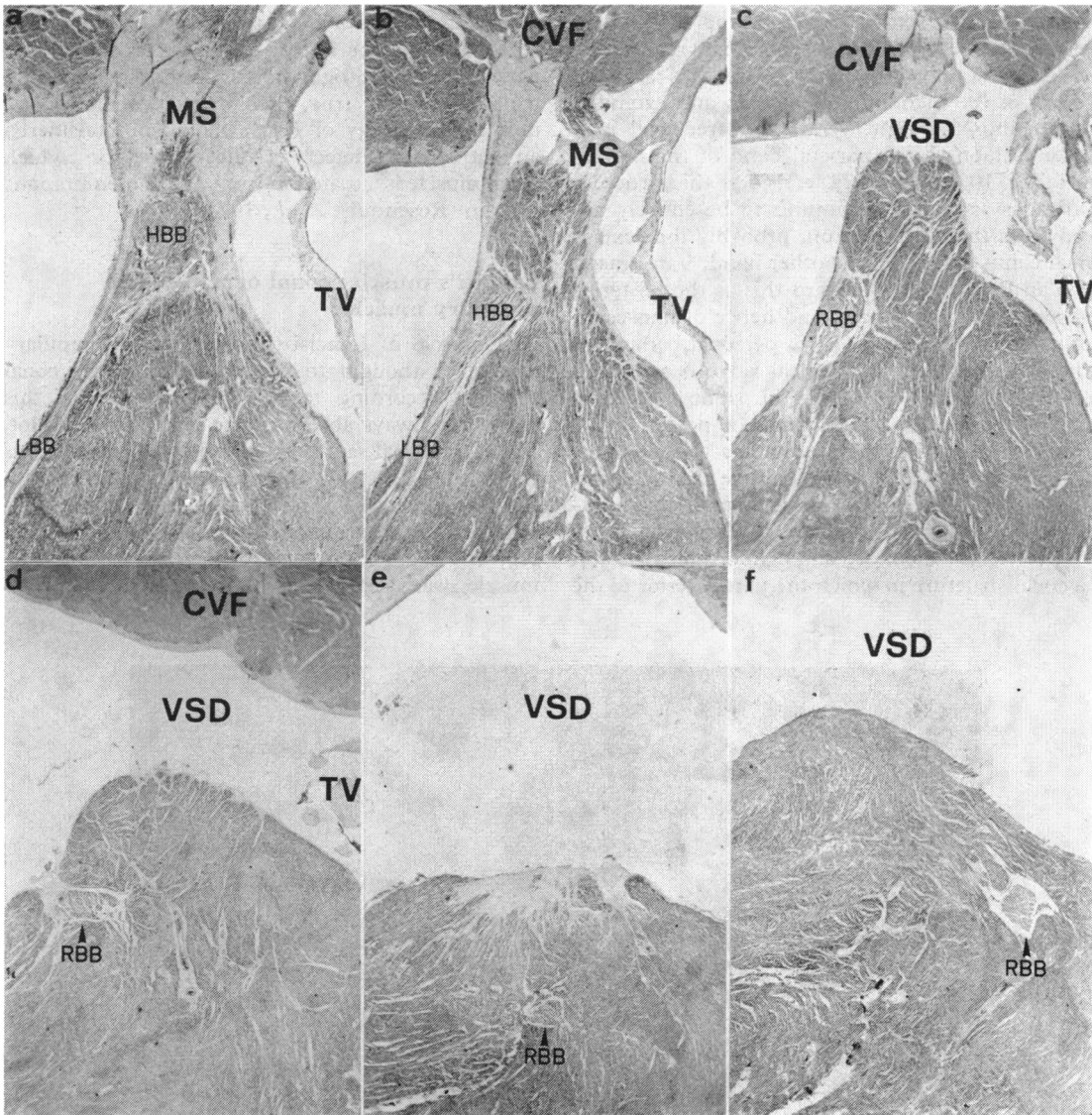
FIG. 8 *Microphotographs of AV conducting system, from case with partially obstructive ventricular septal defect, studied with serial histological sections. Junctional area sectioned in frontal plane from posterior to anterior. (a) and (b) Branching main bundle runs beneath and on left of membranous septum, giving rise to left bundle-branch, then passing to left of crest of posterior muscular septum. In serial sections sheet-like left branch originates before ventricular septal defect appears. (c) First appearance of the ventricular septal defect, posteriorly: left bundle-branch can no longer be seen; right bundle-branch evident on left side of crest of ventricular septum as direct continuation of branching main bundle. (d, e, f) Serial sections in posteroanterior sequence, showing intramyocardial course of cord-like right bundle-branch, which moves from left to right; only right bundle-branch is related to inferior border of ventricular septal defect. CVF=conoventricular flange, HBB=branching His bundle, LBB=left bundle-branch, MS=membranous septum, RBB=right bundle-branch, TV=tricuspid valve, VSD= ventricular septal defect.*

truncal and tricuspid valves appeared in some of these to be a well-formed muscular mass resembling the 'crista supraventricularis'. This was situated in front of the membranous septum and extended laterally into the right ventricular free wall in a similar fashion to the parietal band of the normal heart (Fig. 10). Goor (1972) considers this structure in truncus arteriosus communis to be entirely derived from the conal septum, probably the dextro-dorsal conus ridge. On the other hand, Van Praagh and Van Praagh (1965) regard this as the posterior division of the septal band and hence of non-conal origin. R. H. Anderson (1975, personal communication) believes that the truncal valve is separated from the tricuspid valve by the right margin of the conoventricular flange, while a posterior extension of the septal band separates the membranous septum and the tricuspid valve from the rim of the defect. We hold that, like the crista supraventricularis of the normal heart (Anderson *et al.* 1974b; Becker, Connor, and Anderson, 1975), it is a conal structure in which the part anterior to the

membranous septum is a vestige of the conal septum, while the lateral extension represents the persistence of the right side of the conoventricular flange. If this is true, then the ventricular septal defect in tetralogy of Fallot should be considered an intracristal defect in those cases in which conal muscle is situated in front of the membranous septum (Rosenquist *et al.*, 1973).

Lancisi's muscle: conal or non-conal papillary muscle?

The muscle of Lancisi (or right medial papillary muscle) is thought to be derived from the conal septum; according to Van Mierop (1970), this muscle is always absent in the tetralogy of Fallot, where the conal septum is anteriorly deviated, thus confirming its conal origin. On the contrary, we have identified a Lancisi's muscle in truncus arteriosus communis specimens where no trace of conal septum was present, i.e. those cases where no muscle was seen in front of the membranous
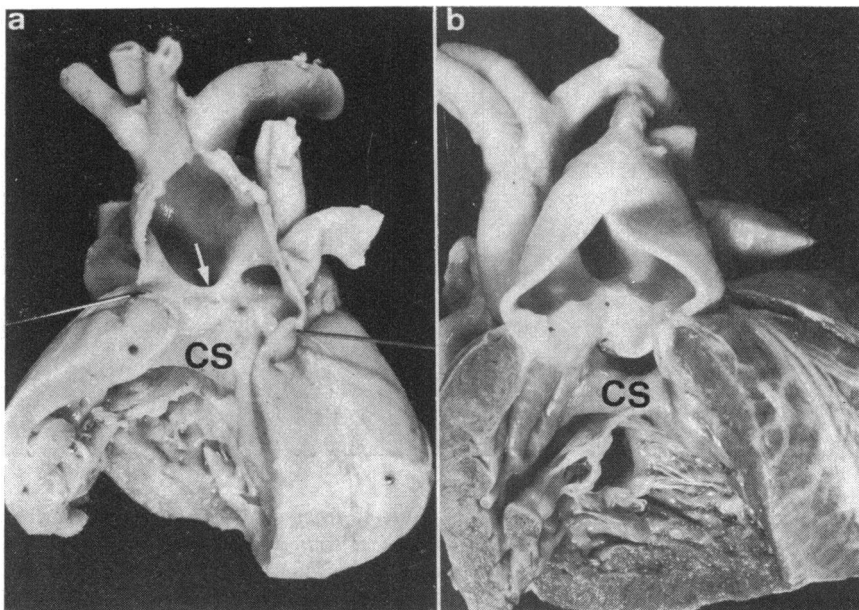
FIG. 9 *Malformations developmentally related to truncus arteriosus communis. (a) Aortopulmonary septal defect: though both infundibula and the semilunar valves are normally divided, a large communication exists between ascending aorta and common pulmonary trunk. Note that the sinus portions of aorta and pulmonary artery are separated by a fibrous septum (white arrow); this probably represents the most cranial extension of the truncal septum and indicates the limit between truncal and the aortopulmonary septa. (b) Isolated supracristal ventricular septal defect: large communication between infundibula of right and left ventricles; semilunar valves, ascending aorta, and pulmonary trunk normally separated. CS = conal septum.*

septum (Fig. 7b). In our opinion, these findings suggest that Lancisi's muscle from a developmental viewpoint is not a 'conal' papillary muscle.

Spectrum of defects of septation in truncus arteriosus communis and related malformations

From the examination of specimens of hearts with truncus arteriosus communis and related malformations, the existence of three different developmental components, taking part in the division of the outflow tracts of the heart, may be postulated (Van Mierop and Gessner, 1972): the conal septum, separating the infundibula; the truncal septum, separating the semilunar valves and the sinus portions of aorta and pulmonary artery; and the aortopulmonary septum separating the ascending aorta from the pulmonary trunk. Though the embryological border between truncus and aortic sac has not yet been defined, our findings suggest that the sinus portion of the great arteries

is derived from the embryological truncus, while the tubular portion is derived from the aortic sac.

In truncus arteriosus communis there is a lack of septation of the truncus and conus, resulting from a failure of development of the truncal and conal septa, while the aortopulmonary septum may (Type I) or may not (Types II, III, IV) have developed. On the other hand, in the cases of aortopulmonary window and conal ventricular septal defect, there is an isolated failure of growth of greater or lesser degree of the aortopulmonary and conal septa, respectively. Thus, in aortopulmonary septal defect truncal and conal septa are usually well formed, with resulting normal separation of the ventricular infundibula and semilunar valves; in a similar manner, in conal ventricular septal defects the aortopulmonary and truncal septa have developed normally.

Fig. 11 shows all the possible malformations which may theoretically occur as a result of single or multiple failures of the three developmental septation components. A malformation featuring
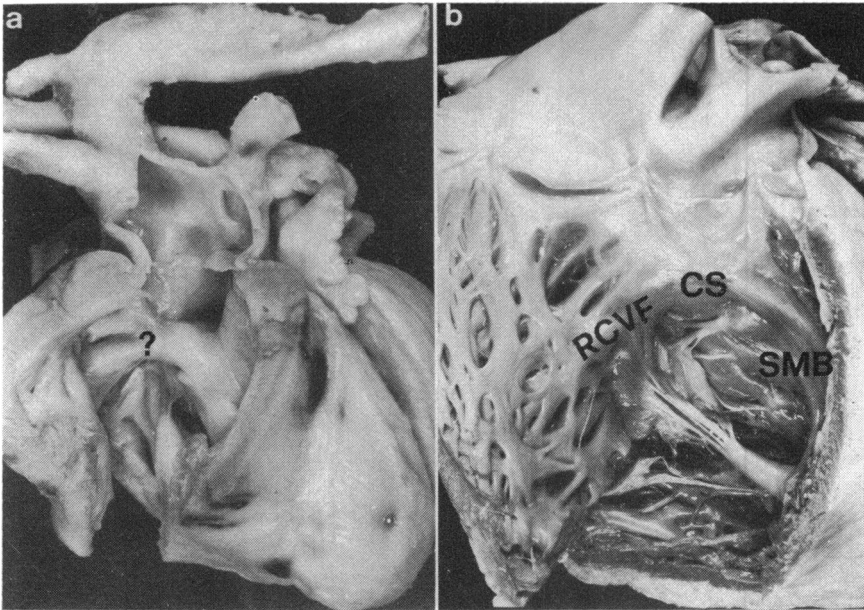

FIG. 10 *(a) Truncus arteriosus—right ventricular outflow tract: question mark indicates thick muscular structure, partly situated in front of membranous septum and extending laterally towards ventricular free wall; Lancisi's muscle is present below that structure. Ventricular septal defect resembles a supracristal ventricular septal defect. (b) c.f. right ventricular outflow tract in normal heart. This muscular mass in truncus is similar to crista supraventricularis of normal heart. Portion of normal crista in front of membranous septum may be vestige of proximal conal septum (CS) in its dextrodorsal conus ridge, while lateral extension or parietal band may be remnant of right portion of conoventricular flange (RCVF); septal extension is septomarginal band (SMB).*

separated ventricular infundibula (normally formed conal septum) and a common semilunar valve (absent truncal septum), with or without aorto-pulmonary septum, has never been reported. In addition, aortopulmonary window might exist together with conal ventricular septal defect; in fact, only membranous ventricular septal defects have been described associated with aortopulmonary window (Tandon *et al.*, 1974). We suggest that these 'missing links' in congenital heart diseases may exist, though examples have not yet been reported.

What is Type IV truncus arteriosus communis?

There is no agreement on whether or not Type IV lesions should be included in truncus arteriosus communis. Macartney, Deverall, and Scott (1973) took the view that Type IV truncus should not be regarded as a form of truncus arteriosus communis; the fourth and sixth aortic arches are not 'communis' since there is agenesis of the latter. Type IV has been called 'solitary aorta associated with absence of the pulmonary arterial system' (Van Praagh and Van Praagh, 1965), 'extreme
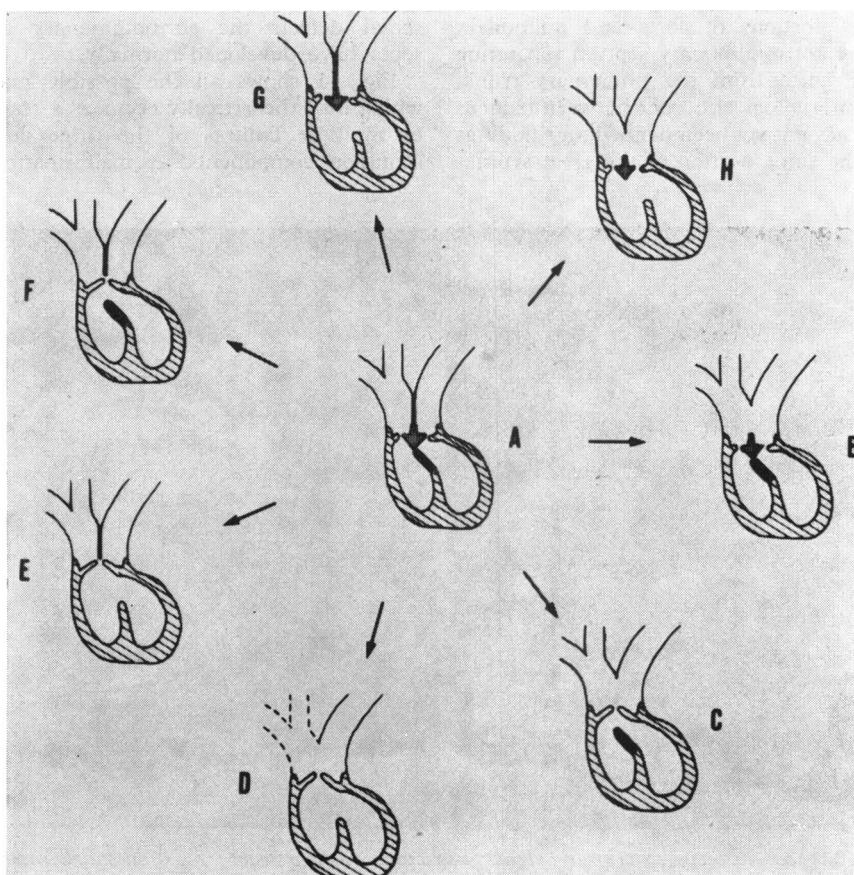

FIG. 11   *Schematic representation of all theoretical malformations derived from single or multiple developmental failure of conal, truncal, and aortopulmonary septa. (A) normal heart; (B) the isolated aortopulmonary septal defect; (C) absent truncal and aortopulmonary septa, with normally developed conal septum (not described so far); (D) Type II, III, IV truncus; (E) Type I truncus; (F) isolated truncal septal failure (not described); (G) isolated conal septal defect; (H) associated aortopulmonary and conal septal defects (not reported). (To simplify the drawings, the various septa are represented as totally present or absent.)*

pulmonary atresia similar to pseudotruncus associated with agenesis of pulmonary artery' (Stuckey, Bowdler, and Reye, 1968; Macartney *et al.*, 1973), and 'true truncus arteriosus communis associated with bilateral agenesis of the sixth aortic arch' (Collett and Edwards, 1949; Bharati *et al.*, 1974). Because of the lack of pulmonary trunk remnants and the absence of a pulmonary infundibulum, we do not consider our cases as examples of extreme tetralogy of Fallot. No signs of truncoconal septation were present, indicating a complete failure of partitioning of the conus and truncus, as in the other types of truncus arteriosus communis. Accepting the hypothesis that Type IV is a true truncus arteriosus communis, we consider that the absence of the sixth aortic arches must be regarded as an additional developmental deficiency in these cases. Thus, in Type IV truncus there is a single, undivided conotruncus, associated with an agenesis of the sixth aortic arches.

There are conflicting views regarding the nature of the arteries supplying the lungs in Type IV truncus. In the case where the left pulmonary circulation was demonstrated *post mortem*, the systemic arteries to the lungs arose from the descending aorta, entered the hila, and anastomosed directly with the arteries normally branching in the parenchyma. For this reason, we cannot regard these as true bronchial arteries, a view shared by Macartney *et al.* (1973). We believe that this systemic blood supply to the lungs in Type IV truncus is derived from a direct embryological vascular connexion between the dorsal aorta and the pulmonary plexus, as an alternative to the undeveloped sixth aortic arches. According to Jefferson, Rees and Somerville (1972), these vessels may be derived from persistent intersegmental branches of the dorsal aorta which represent the primitive blood supply to the lungs in early stages of development (Boyden, 1970). If the ventral portion of the sixth aortic arch is absent and the dorsal portion develops, the systemic circulation to the lungs in Type IV truncus may be supplied by bilateral persistent ductus arteriosus, as described by Murray and co-authors (1970).

Underdevelopment of aortic arch in truncus arteriosus communis

Hypoplasia or interruption of the aortic arch was observed in 3 of our cases, and in 14·4 per cent of cases in the extensive necropsy series recently reported by Bharati *et al.* (1974). In our specimens, the underdevelopment of the aortic arch was also associated with a widely patent ductus arteriosus, confirming the rule that there is an inverse relation

between the development of the fourth and sixth aortic arches in truncus arteriosus communis (Van Praagh and Van Praagh, 1965). As suggested by Van Mierop (1970), the fetal persistence of both these arches in truncus arteriosus communis is superfluous, because the ventricular outflow tracts and the ventral aorta are not divided and two connexions with the descending aorta are not necessary. We have emphasized (Thiene, Cucchini, and Pellegrino, 1975) that underdevelopment of the aortic arch in truncus arteriosus communis seems to be constantly associated with a univentricular origin of the truncus from the right ventricle, suggesting that the right lateral displacement of the truncus results in an uneven distribution of the common blood flow between the ascending aorta and the pulmonary artery; consequently, the predominant flow is from right to left through the ductus arteriosus, predisposing to the involution of the fourth aortic arch. Origin of the truncus arteriosus communis from the right ventricle was present in all our three cases, and also in those reported cases in which the alignment of the truncus with the ventricles was described in detail (Newcombe *et al.*, 1961; Steiner and Finegold, 1965). This association warrants further study and confirmation.

Surgical considerations

The transformation of the truncus and truncal valve, respectively, into aorta and aortic valve and the incorporation of the ventricular septal defect into the left ventricular outflow tract, represent the main steps in the total correction of truncus arteriosus communis (McGoon *et al.*, 1968). Consequently, associated anomalies of the aorta, of the truncal valve, and of the ventricular septal defect may create additional problems for the surgeon.

Origin of the truncus entirely from the right ventricle implies that the ventricular septal defect, as a bulboventricular foramen, functions as the only outlet for the left ventricle. If the size of the defect is inadequate, thus obstructing the left ventricular outflow, surgical enlargement must be considered while planning the radical operation; under these circumstances, knowledge of the location of the conducting tissue is important for the surgeon. In the specimen studied by serial histological sections, the His bundle ran posterior to the ventricular septal defect and the left bundle-branch originated from the branching bundle before reaching the ventricular septal defect. Apart from the aberrant course of the right bundle-branch, these findings are similar to those described

in hearts with Fallot's tetralogy or isolated membranous ventricular septal defects (Truex and Bishof, 1958; Lev, 1959; Visioli, Baragan, and Lenègre, 1962; Titus, Daugherty, and Edwards, 1963; Latham and Anderson, 1972), in which the perforating and the branching positions of the AV bundle were related to the posteroinferior margin of the septal defect. From a surgical point of view this implies that the main bundle and the left bundle-branch are in a 'safe' position, while only the right bundle-branch seems to occupy the area at risk in the inferior border of the ventricular septal defect. Latham and Anderson (1972) have documented pathologically a case in which intraoperative trauma to the right bundle-branch produced a haematoma which tracked back to the branching AV bundle and caused heart block. Thus, if enlargement of the ventricular septal defect is needed in truncus arteriosus communis, the manoeuvre should involve only its anterosuperior quadrant or the conoventricular flange if present, with avoidance of trauma to the inferior border of the defect.

Interruption or tubular hypoplasia of the aortic arch represents a severe additional lesion. So far, only 3 cases of total repair of truncus arteriosus communis associated with interrupted aortic arch have been reported (Mair *et al.*, 1974). The additional association of origin of the truncus from the right ventricle in these cases complicates the problem further, especially if the ventricular septal defect needs to be enlarged.

The truncal valve was abnormal in most of our specimens, and abnormal function of the truncal valve has also been frequently described in clinicopathological reports (Tandon, Hauck, and Nadas, 1963; Deely, Hagstrom, and Engle, 1963; Victorica *et al.*, 1969; Burnell, McEnery, and Miller, 1971; Gelband, Van Meter, and Gersony, 1972). In addition to those cases of gross dysfunction, which may require valve replacement at the time of total correction, increasing the surgical risk, the long-term effects of minor abnormalities should be carefully evaluated. One of our patients, a 29-year-old man, presented with a moderate truncal valve stenosis and nodular calcification of the cusps. As suggested by Becker *et al.* (1971), nodular thickening of the truncal valve may be an acquired lesion resulting from haemodynamic stress in the presence of a pre-existing developmental malformation. The state of the truncal valve should be taken into account when assessing the prognosis after operation in these cases.

Reconstruction of continuity between the right ventricle and the aortopulmonary collateral vessels might theoretically be indicated in Type IV cases, because of the direct anastomoses between the thoracic aorta and the pulmonary arterial bed. However, the site of origin, the multiplicity of the vessels, and the frequent stenoses at the lung hila precluded surgery in almost every case (Macartney *et al.*, 1973; Chesler, Matisonn, and Beck, 1974); to our knowledge, only one such patient has been successfully operated (McGoon, Wallace, and Danielson, 1973). However, the patient whose heart is shown in Fig. 2, who had a large left-to-right shunt causing lethal left ventricular failure, could have been an ideal candidate for correction. We agree with Chesler *et al.* (1974) that direct selective catheterization and arteriography of these systemic collateral vessels is essential in the investigation of these patients for operation.

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