Pulmonary atresia with ventricular septal defect *Further anatomical observations*

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An anatomical study was undertaken of 25 hearts with pulmonary atresia and ventricular septal defect, giving special attention to the morphology of the pulmonary outflow tract and the nature of the arterial supply to the lungs. A common feature of all hearts studied was the presence of sixth aortic arch derivatives within the pericardial cavity. Eighteen specimens presented with pulmonary atresia with ventricular septal defect in situs solitus, with atrioventricular concordance and normally related great arteries, that is with the pulmonary artery remnant anterior and to the left of the aorta. A blind or atretic pulmonary infundibulum was observed in all but one of these cases. In the remaining 7 specimens, the atretic pulmonary trunk was posterior to the aorta.

The pulmonary arterial blood supply was either through a ductus arteriosus or systemic-pulmonary anastomotic vessels. Anastomotic vessels were never observed together with a ductus, suggesting that the systemic-pulmonary vessels represent persistence of early arterial connections with the lungs when the ductus fails to develop.

The result of this study when compared with our previous studies indicates that a distinction can be made between pulmonary atresia with ventricular septal defect and persistent truncus arteriosus with absent pulmonary arteries ('type IV'). The latter is considered to be a true truncus arteriosus in which the arterial connections with the pulmonary vascular bed through the sixth aortic arches failed to develop, lack of septation of the cardiac outflow tracts rendering them superfluous, and thus only the early systemic-pulmonary connections persist. However, the condition is difficult or impossible to distinguish from pulmonary atresia with ventricular septal defect on clinical grounds.

In the examples of pulmonary atresia with ventricular septal defect and posteriorly placed pulmonary trunk remnants, the pulmonary circulation was always supplied by the ductus arteriosus; systemic-pulmonary arteries were never seen. The mechanism producing atresia of the posteriorly positioned pulmonary infundibulum seems to be different from the form with anteriorly placed pulmonary infundibulum, as no deviation of the conal septum was seen and the ventricular septal defect was rarely a malalignment defect.

Conflicting opinions exist concerning the classification, terminology, and precise nature of pulmonary atresia with ventricular septal defect. The wide variety of terms used to describe the malformation reflects this discordance of views as to its nature. Thus the anomaly has been termed truncus solitarius aorticus (Kugel, 1931), pulmonary atresia with biventricular aorta (Muñoz-Armas *et al.*, 1964), pseudotruncus arteriosus (Edwards *et al.*, 1965; Chesler *et al.*, 1970), extreme tetralogy of Fallot (Stuckey *et al.*, 1968), truncus arteriosus type IV (Taussig *et al.*, 1973), or has been described as

single arterial trunk, aorta (Shinebourne *et al.*, 1976).

Since many examples of the anomaly have now undergone correction, it has been the subject of numerous clinical (Miller *et al.*, 1968; Somerville, 1970; Danilowicz and Ross, 1971; Jefferson *et al.*, 1972; Macartney *et al.*, 1973; Chesler *et al.*, 1974; Macartney *et al.*, 1974) and surgical reports (Ross and Somerville, 1966; McGoon *et al.*, 1970; Kouchoukos *et al.*, 1971; Doty *et al.*, 1972; Somerville and Ross, 1972; Berry *et al.*, 1974; Pacifico *et al.*, 1974; McGoon *et al.*, 1975; Marcelletti *et al.*, 1976). In contrast, few pathological investigations

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have been published (Muñoz-Armas *et al.*, 1964; Edwards and McGoon, 1973; Bharati *et al.*, 1975). It seemed that the anatomy of this cardiac defect had not yet been satisfactorily described. The present study was undertaken, specifically, to clarify the morphology of the ventricular outflow tracts and the arterial supply to the lungs.

Definition of terms

We define pulmonary atresia with ventricular septal defect as a malformation in which both ventricles eject into the aorta and in which some derivatives of the sixth aortic arches can be identified inside the pericardial cavity. When such derivatives are absent, we believe the anomaly to be a variant of persistent truncus arteriosus (Thiene *et al.*, 1976). In pulmonary atresia with ventricular septal defect there is a lack of continuity between the heart and the pulmonary artery and blood is supplied to the lungs by vessels derived from the aortic arch and the descending aorta. Twenty-five hearts from our collection showed these features.

Cases of pulmonary atresia with intact ventricular septum, atresia of an atrioventricular valve, common atrioventricular orifice, or primitive ventricle were excluded from this review.

The term 'transposition of the great arteries' has not been used throughout the paper to describe cases with anterior aorta and posterior atretic pulmonary trunk, because in this condition it is often impossible to determine if the pulmonary artery has indeed been placed across the septum (Shinebourne *et al.*, 1976).

Subjects and methods

The 25 anatomical specimens have been classified according to: viscerotrial situs; atrioventricular connections; cardiac position; and the spatial relation of the great arteries.

(a) SITUS SOLITUS (22 cases)

Eighteen cases presented with the classical features of pulmonary atresia with ventricular septal defect: laevocardia, concordant atrioventricular connection, and normally related great arteries, i.e. the atretic pulmonary artery was anterior and to the left of the aorta and potentially connected to the right ventricle. In contrast, 2 cases showed laevocardia, concordant atrioventricular connection, and single arterial trunk, but the atretic pulmonary artery was in a posterior position, being to the left of the aorta. The 2 remaining cases had the atretic pulmonary artery in the posterior position, but to the right of the aorta in association with atrioventricular dis-

cordance, one with laevocardia and the other with dextrocardia.

(b) SITUS INVERSUS (3 CASES)

Two hearts showed laevocardia, discordant atrioventricular connection, a posterior blind-ending pulmonary trunk, and an aorta to the right, while one presented with dextrocardia, concordant atrioventricular connection, a posterior blind-ending pulmonary artery, and an aorta to the left.

Fig. 1 Morphology of the right ventricular outflow tract in two examples of pulmonary atresia with ventricular septal defect and normally related great arteries. (a) The pulmonary infundibulum (PI) is short and blind and the conal septum (CS) is anteriorly deviated and inserted behind the trabecula septomarginalis (TSM). (b) The pulmonary infundibulum is atretic from its origin.

Fig. 2 Morphology of the right ventricular outflow tract in pulmonary atresia with ventricular septal defect and no identifiable pulmonary trunk. (a) No pulmonary trunk is identifiable while a left ductus arteriosus (LDA) is connected with two confluent pulmonary arteries. The aortic arch is on the right side. (b) Right ventricular view: the conal septum (CS) is a distinct muscular structure fused with the base of the right ventricular free wall and inserted in front of the trabecula septomarginalis (TSM). The pulmonary infundibulum is completely atretic from its origin.

All hearts were examined with specific attention to the morphology of the ventricular outflow tracts and the ventricular septal defect, arterioventricular alignment and aortic arch, coronary arterial patterns, systemic-pulmonary arterial supply, and size of the pulmonary trunk and its main branches.

Findings

After examining the hearts, the specimens were divided into 2 groups, on the basis of the *relation* of the great arteries, it being implicit in our definition that the ventriculoarterial connection was one of single arterial trunk.

Group 1 had the blind-ending pulmonary infundibulum or atretic pulmonary artery in the anterior position, so that the arterial relations were normal. Group 2, in contrast, had a posteriorly situated atretic pulmonary trunk.

GROUP 1: PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND NORMALLY RELATED GREAT ARTERIES (18 CASES)

(a) Morphology of ventricular outflow tracts and ventricular septal defect

In 13 hearts the pulmonary infundibulum was severely hypoplastic, anterior, and short, and the atresia was situated at valvular level (Fig. 1a). In 4 cases the pulmonary infundibulum appeared atretic from its origin: nevertheless the conal septum was identified as a distinct muscular structure fused with the anterior free wall of the right ventricle and the ventricular septum, in front of the trabecula septomarginalis (Fig. 1b). In one of these specimens, despite careful gross and histological examination, no pulmonary trunk was recognisable, while 2 confluent and moderately developed pulmonary arteries were supplied by a left ductus arteriosus (Fig. 2). In only one case (Fig. 3a), were we unable to identify either a blind or a potential pulmonary infundibulum as a separate entity from the sinus portion of the right ventricle. In this heart, no trace of the conal septum was found below the root of the great arteries. Nevertheless, a thin, fibrous, cordlike structure connected the heart with two confluent and relatively large pulmonary arteries into which a wide left ductus arteriosus opened. On histology, the fibrous cord was shown to be a vascular structure with a lumen and an elastic medial layer, and thus it was interpreted as a pulmonary trunk (Fig. 3b).

The anterior septal leaflet of the tricuspid valve was in fibrous continuity with the posterior aortic cusp in all but one case in which a small muscular band was interposed between these structures.

The left ventricular outflow tract always showed mitral-aortic fibrous continuity.

The ventricular septal defect was inferior and posterior to the deviated conal septum and derived from the aortic overriding and from the malalignment between the conal and ventricular septa, as described in specimens of tetralogy of Fallot (Becker *et al.*, 1975).

The membranous septum was partially formed, constituting the posterior rim of the ventricular septal defect in all but one case. In two instances, this partially formed interventricular component was sufficiently redundant to produce some occlusion of the right ventricular outflow to the aorta (Fig. 4).

Fig. 3 Morphology of the right ventricular outflow tract in pulmonary atresia with ventricular septal defect and no macroscopically recognisable pulmonary trunk or infundibulum. (a) The right ventricular outflow tract is similar to that described in truncus arteriosus communis without remnants of the conal septum. (b) Histology of the root of the aorta (A) shows a small vascular structure, with a lumen and an elastic medial layer, which represents the main pulmonary artery (PA). (Elastic van Gieson stain. $\times 12$.)

(b) Arterioventricular alignment and aortic arch

The aorta arose from the right ventricle to an extent which varied between 50 and 100 per cent. There was no relation between the degree of aortic overriding and the underdevelopment of the pulmonary infundibulum. The aortic arch was left-sided in 13 cases and right-sided in 5; 4 of the latter had a left ductus arteriosus.

(c) Coronary arterial patterns

The aortic root, as viewed from the ventricular aspect, was rotated clockwise so that the right coronary cusp was displaced anteriorly. In all instances the coronary arteries arose from the 2

sinuses adjacent to the atretic infundibulum. In 2 cases the epicardial course of the coronary arteries was abnormal in that the anterior descending branch arose from the right coronary artery and crossed the right ventricular infundibulum to reach the septum.

(d) Systemic-pulmonary arterial supply, size of pulmonary trunk, and of its main branches

The right and left pulmonary arteries were always confluent. In 7 specimens a ductus arteriosus was persistent, the pulmonary trunk was hypoplastic or rudimentary (absent in one case) and the pulmonary arteries, though well developed, were

Fig. 4 Fibrous tissue (arrows) from membranous septum and septal leaflet of the tricuspid valve is so redundant that the aortic outflow tract from the right ventricle is obstructed. CS, conal septum; PI, pulmonary infundibulum. (a) Right ventricular view; (b) left ventricular view.

Fig. 5 Pulmonary arterial supply in 3 examples of pulmonary atresia with ventricular septal defect and normally related great arteries. (a) A left sided ductus arteriosus (LDA) connects a left aortic isthmus with the left pulmonary artery. The pulmonary trunk (PT) is hypoplastic and the pulmonary arteries smaller than normal. (b) A large left ductus arteriosus (LDA) is the only source of pulmonary arterial supply as in case (a). Here the pulmonary trunk and arteries are normal in size. (c) Agenesis of the ductus arteriosus. The pulmonary arterial supply is furnished by 3 systemic pulmonary arteries (SA), 2 for the left lung and 1 for the right lung: the 3 vessels do not connect with the major pulmonary arteries (PA). The pulmonary trunk and arteries are extremely hypoplastic.

smaller than normal. No systemic-pulmonary anastomotic vessels were observed in these cases (Fig. 5a).

Three specimens showed a good sized pulmonary trunk and pulmonary arteries with a wide ductus arteriosus and no other systemic-pulmonary arterial supply (Fig. 5b).

In 7 other specimens the ductus was absent and the pulmonary trunk and its main branches were severely hypoplastic. Small systemic-pulmonary arteries, originating from the descending aorta, entered both lungs without anastomosing with the major pulmonary arteries at the hilum (Fig. 5c).

Finally, in one case, even though the ductus was absent, the pulmonary trunk and the pulmonary arteries were of reasonable size, because of the existence of a direct anastomosis at the hilum of the

right lung, between the right pulmonary artery and a large systemic-pulmonary artery arising from the descending aorta (Fig. 6). In the same patient other large systemic-pulmonary arteries entered both lungs without anastomosing with the proximal pulmonary vascular bed (Fig. 6b). This patient, who died of left ventricular failure at age 9 months without undergoing operation, was acyanotic. Histological examination of his lungs disclosed mild medial hypertrophy of the small pulmonary arteries and arterioles (Fig. 6d).

GROUP 2: PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND POSTERIORLY SITUATED PULMONARY TRUNK (7 CASES)

The morphological features of this group may be summarised as follows (Fig. 7).

Fig. 6 Pulmonary arterial supply in a case of pulmonary atresia with ventricular septal defect and normally related great arteries in which the ductus was absent. (a) Anterior external view: though the ductus arteriosus is absent the pulmonary trunk and arteries are normally sized. (b) Posterior view (oesophagus and trachea removed): four large systemic-pulmonary arteries (SA) arise from the descending aorta, one for the left lung and three for the right lung. (c) One of these large systemic arteries (SA) anastomoses with the right pulmonary artery (RPA) at the hilum of the right lung. (d) Histology of the lungs reveals mild medial hypertrophy and duplication of the elastic layers in the small muscular pulmonary arteries. (Elastic van Gieson stain. $\times 150$.)

(a) The aortic root arose entirely from the right ventricle without overriding the left (Fig. 7a), except in one case when there was 50 per cent override (Fig. 7b).

(b) The subaortic conus showed a muscular band (conoventricular flange) interposed between the aortic and the atrioventricular valves.

(c) The ventricular septal defect derived from a partial or complete absence of conal septum and was situated below it.

(d) In all hearts the pulmonary circulation was

supported only by a ductus arteriosus with moderately-sized pulmonary arteries, while systemic-pulmonary anastomotic vessels were never seen.

Discussion

PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND NORMALLY RELATED GREAT ARTERIES

The present findings and our previous observations (Thiene *et al.*, 1976) constitute strong evidence that

Fig. 7 Right ventricular outflow tract morphology and types of ventricular septal defect in pulmonary atresia with ventricular septal defect and posterior atretic pulmonary artery.

(a) The aorta arises entirely from the right ventricle above a muscular conus. The ventricular septal defect (VSD) is an intracristal defect and the conal and ventricular septa are normally aligned (from a patient with situs inversus, laevocardia and atrioventricular discordance). CS, conal septum; CVF, conoventricular flange.

(b) The aorta presents with biventricular origin and the ventricular septal defect, which lies between the anterior and posterior limbs of the trabecula septomarginalis (TSM), derives from the absence of the conal septum (from a patient with situs inversus, dextrocardia, and atrioventricular concordance).

Fig. 8 The spectrum of pulmonary outflow obstruction in normally related great arteries—(a) normal heart, (b) tetralogy of Fallot, (c) pulmonary atresia with ventricular septal defect. Note the progressive anterior deviation of the conal septum with its septal insertion in front of the trabecula septomarginalis. CS, conal septum; CVF, conoventricular flange; PI, pulmonary infundibulum; TSM, trabecula septomarginalis.

the malformation of pulmonary atresia with ventricular septal defect and persistent truncus arteriosus is quite distinct, both morphologically and morphogenetically. In truncus arteriosus the septation of the cardiac outflow tracts is absent, thus resulting in a conal ventricular septal defect and a common semilunar valve. In contrast, in pulmonary atresia with ventricular septal defect unequal septation occurs at the expense of the pulmonary infundibulum, and the ventricular septal defect results from the aortic overriding and from the extreme malalignment between the anteriorly deviated conal septum and the interventricular septum (Goor *et al.*, 1971; Becker *et al.*, 1975). We, therefore, believe that it is confusing and misleading to group under the term 'absence of anatomic origin from the heart of pulmonary arterial supply' such dissimilar cardiac malformations as truncus arteriosus and pulmonary atresia even though both are characterised by discontinuity between the right ventricle and the pulmonary artery (McGoon *et al.*, 1970; Edwards and McGoon, 1973; Berry *et al.*, 1974). We also believe that our evidence militates against the concept proposed by Van Praagh that truncus arteriosus should be considered as extreme pulmonary atresia with ventricular septal defect and an aortopulmonary septal defect (Van Praagh and Van Praagh, 1965; Calder *et al.*, 1976; Van Praagh, 1976). Thus, in all our cases of pulmonary atresia with ventricular septal defect, derivatives of the sixth aortic arches were observed in the pericardial cavity and in all but one a pulmonary trunk arose from the heart base suggesting truncal septation. In the single specimen in which a main pulmonary artery could not be identified, even by histology, the intracardiac anatomy of the right ventricle showed an atretic pulmonary infundibulum with a distinct conal septum remnant. Thus in all our specimens signs of conal, truncal, or truncoconal septation were present. These findings indicate that pulmonary atresia should be included in the spectrum of tetralogy of Fallot as it is characterised by the same, albeit extreme, malseptation, i.e. conal rotation and anterior deviation of the conus septum which determine, respectively, dextroposition of the aorta and atresia of the pulmonary infundibulum (Fig. 8). Furthermore, pathological evidence shows that pulmonary atresia with ventricular septal defect and absent ductus arteriosus participates in the spectrum of Fallot's tetralogy in Keeshond dogs with hereditary defects of the conotruncal septum (Patterson *et al.*, 1974). In contrast, much of the evidence at present available shows that persistent truncus arteriosus results from failure of septation of the embryonic infundibula and truncus (Thiene *et al.*, 1976; Crupi *et al.*, 1977).

PULMONARY CIRCULATION IN PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND NORMALLY RELATED GREAT ARTERIES

In pulmonary atresia with ventricular septal defect, unlike truncus arteriosus, the pulmonary circulation does not originate directly from the ascending aorta. The alternative sources of pulmonary blood supply are either the ductus arteriosus or systemic-pulmonary anastomotic vessels which arise from the descending aorta.

In 38 per cent of our cases the ductus arteriosus was absent, thus determining extreme underdevelopment of the pulmonary arteries. Under these conditions the pulmonary arterial supply is furnished only by systemic vessels which anastomose intraparenchymally with the intrapulmonary arterial bed. In a single specimen presenting with absent ductus, the pulmonary trunk and its branches were wide because of a proximal anastomosis at the hilum between the right pulmonary artery and a large systemic vessel. On the other hand, in the 10 cases where a ductus arteriosus was present, the pulmonary arteries had a diameter ranging from half normal to normal size. Our findings are in agreement with those of Bharati *et al.* (1975) regarding the inverse relation between the presence of a ductus arteriosus and the presence of systemic-pulmonary arteries. Angiographic investigations have also shown that systemic-pulmonary arteries and ductus arteriosus do not coexist (Macartney *et al.*, 1974).

According to Jefferson *et al.* (1972), there are two basically different types of collateral vessels, namely large and small. Macartney *et al.* (1974) argued that the small type might be acquired, but our findings of small systemic-pulmonary anastomotic vessels in specimens from newborn patients lead us to disagree with this deduction.

Various terms have been suggested to describe these anastomotic vessels: bronchial arteries (Chesler *et al.*, 1974), aortico-pulmonary collateral arteries (Kouchoukos *et al.*, 1971; Macartney *et al.*, 1974), and systemic arteries (Jefferson *et al.*, 1972). The term 'bronchial arteries' is inaccurate from both an anatomical and embryogenetic point of view. The arteries may take origin from brachiocephalic arteries or subdiaphragmatic aorta as well as from the thoracic aorta (McGoon *et al.*, 1975). Moreover, these arteries derive from the primitive intersegmental branches of the dorsal aortae which supply the lung bud early in embryonic life (Congdon, 1922; Boyden, 1970), and we believe that these arteries should be called systemic-pulmonary arteries. The persistence of these early embryological connections between the systemic

and pulmonary circulations occurs in pulmonary atresia whenever the distal sixth aortic arch (ductus arteriosus) fails to develop. Furthermore, this failure may occur in other situations. Thus absence of septation of the cardiac outflow tracts in truncus arteriosus may render superfluous the growth of the sixth aortic arches as a second connection between the ventral and dorsal aortae, thus determining the persistence of intersegmental arteries as the only source of pulmonary blood supply, such as seen in persistent truncus arteriosus with absent pulmonary arteries ('type IV') (Table).

PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT VERSUS TRUNCUS ARTERIOSUS TYPE IV

Taussig *et al.* (1973) have suggested that 'pseudo' truncus arteriosus is not a variant of tetralogy of Fallot and that a 'pseudo' truncus arteriosus and a truncus arteriosus type IV are basically the same malformation even though in the former a rudimentary pulmonary artery can be shown at necropsy either through gross or microscopical examination. In similar fashion, Stuckey *et al.* (1968) applied the name 'absent sixth aortic arch' to cases in which derivatives of the sixth aortic arches were present though completely atretic.

It seems to us that these statements add further confusion to an already confused topic. In our opinion it is vital to distinguish between agenesis and atresia of an artery. Thus, the finding of a fibrous strand connecting the heart base to the lungs is incompatible with absence or agenesis of the sixth aortic arches. The fundamental question must, therefore, be as to whether or not the sixth arches have developed. In truncus arteriosus with absent pulmonary arteries ('type IV'), they have not developed, since no remnants of pulmonary arteries are observed (Thiene *et al.*, 1976). In contrast, in

pulmonary atresia with ventricular septal defect, at the very least a fibrous cordlike remnant of the sixth arch is seen within the pericardial cavity, as described in this study. Clearly, at times this distinction may be difficult if not impossible to reach by means of clinical investigation (Jefferson *et al.*, 1972; Macartney *et al.*, 1973; Chesler *et al.*, 1974). None the less, we believe that the distinction should be maintained, since the two malformations are developmentally and morphologically distinct.

Other authors (Somerville, 1970; Macartney *et al.*, 1974) have considered truncus arteriosus 'type IV' as an extreme form of atresia of the pulmonary infundibulum associated with agenesis of the pulmonary arteries. While cases of pulmonary atresia without a pulmonary infundibulum have been reported (Allanby *et al.*, 1950; Chesler *et al.*, 1974), no specimen of complete agenesis of the main pulmonary trunk and pulmonary arteries which exhibits a blind or potential pulmonary infundibulum has thus far been described. It could be argued that truncus arteriosus 'type IV' should not be regarded as truncus arteriosus 'communis' since the absent pulmonary arteries are not contributing to the single arterial trunk. However, if the term 'persistent' instead of 'communis' is used to indicate the truncus arteriosus, as at present, then the anomaly of truncus arteriosus 'type IV' can correctly be termed persistent truncus arteriosus with absent pulmonary arteries, since the anomaly, in addition to exhibiting agenesis of the sixth aortic arches, also possesses a single undivided conotruncus (Thiene *et al.*, 1976).

DIAGNOSTIC AND SURGICAL CONSIDERATIONS

It is difficult to distinguish angiographically between persistent truncus with absent pulmonary arteries and pulmonary atresia with ventricular septal defect when no blind pulmonary infundibulum can be seen and the ductus arteriosus or proximal connections between systemic pulmonary vessels and pulmonary arteries are absent (Jefferson *et al.*, 1972; Chesler *et al.*, 1974; Macartney *et al.*, 1974). The distribution pattern of the systemic-pulmonary arteries is often similar in both conditions; therefore, only direct morphological or surgical observation may establish the presence or absence of sixth aortic arch derivatives, though some authors have argued that it may be expedient to ignore these differences (Crupi *et al.*, 1977).

It is significant that the angiographic features of pulmonary atresia with obstructed ventricular septal defect, secondary to the presence of redundant tissue of the membranous septum, may resemble those of pulmonary atresia with

Table

Defect	Embryology
Truncus arteriosus communis	Absent truncoconal septation with or without distal 6th aortic arch
Persistent truncus arteriosus type IV	Absent truncoconal septation + agenesis 6th aortic arches + persistence of early systemic-pulmonary connections
Pulmonary atresia with ventricular septal defect and without ductus arteriosus	Extreme unequal truncoconal septation + agenesis of distal 6th aortic arches + persistence of early systemic-pulmonary connections
Pulmonary atresia with ventricular septal defect and with ductus arteriosus	Extreme unequal truncoconal septation with normal development of 6th aortic arches

intact ventricular septum and reversed intra-atrial shunt (Scott *et al.*, 1976). It is vital that the two be distinguished, since the prognosis of the two anomalies differs considerably.

Radical surgical correction of pulmonary atresia with ventricular septal defect may be considered only if the right and left pulmonary arteries are well developed (Berry *et al.*, 1974). On this basis, in 7 of our 18 hearts (39%) the pulmonary arteries were too hypoplastic to be used even for palliative surgery.

The systemic-pulmonary arteries theoretically could be used to reconstruct the continuity between the heart and the pulmonary arterial bed. However, their small size and their multiplicity, together with their frequent intrahilar stenoses (Macartney *et al.*, 1973; Chesler *et al.*, 1974) and extrapericardial location make them, for the most part, unsuitable for surgery.

Should it be possible to perform reconstructive surgery, then all of the systemic-pulmonary vessels must be ligated in order to prevent a persistent left-to-right shunt postoperatively (McGoon *et al.*, 1975). Pulmonary overperfusion may be a feature of the disease whenever large and multiple systemic-pulmonary arteries anastomose proximally with the pulmonary arteries (Stuckey *et al.*, 1968; Danilowicz and Ross, 1971).

The finding of specimens where the anterior descending coronary artery, arising from the right coronary artery, crossed the right ventricular infundibulum, confirms previous observations on coronary arterial patterns in pulmonary atresia (Bharati *et al.*, 1975) and Fallot's tetralogy (Fellows *et al.*, 1975). This anomaly can represent an additional surgical difficulty but it might be possible to determine this preoperatively by coronary arteriography.

PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT AND POSTERIOR ATRETIC PULMONARY ARTERY

The condition is characterised by discontinuity between the left ventricle and the pulmonary arterial bed. Surgical repair has been performed also in this variety of pulmonary atresia with ventricular septal defect (Marcelletti *et al.*, 1976).

In all our specimens the source of the pulmonary blood supply was the ductus arteriosus, and systemic-pulmonary arteries were not seen. Moreover, the ventricular septal defect was never a conal malalignment defect, as is the case in pulmonary atresia with normally related great arteries, because the aorta rarely presents with biventricular origin. A ventricular septal defect without malalignment has been previously described in this type of pul-

monary atresia (Kugel, 1931; Young and Griswold, 1951; Kelsey *et al.*, 1953) but its morphogenetic significance has never been underlined. In fact, the mechanism determining atresia of the pulmonary outflow tract in this condition does not seem to depend on the deviation of the conal septum as much as it does in the form with normally related great arteries.

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