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Anatomic Review

Interruption of the Aortic Arch with Aorticopulmonary Septal Defect

An Anatomic Review

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SUMMARY. **Interruption of the aortic arch may occur with aorticopulmonary septal defect (APSD) as part of a developmental complex. The anatomic details of 46 cases of interruption of the aortic arch with APSD revealed the following characteristics: (1) type A interruption of the aortic arch occurred nearly six times more commonly than type B interruption; (2) the APSD was variable in size and position, conforming to the types previously described; (3) the ventricular septum was usually intact, but a ventricular septal defect was present in six cases and occurred more frequently with type B interruption of the aortic arch; (4) subaortic stenosis was not present in any of the 16 cases in which adequate details were available to make a judgment.**
The association of APSD with interruption of the aortic arch in infancy is not uncommon. Evaluation of infants with APSD should thus include evaluation of the aortic arch. The occurrence of interruption of the aortic arch with an intact ventricular septum is distinctly unusual. In such cases an APSD should be suspected and sought.

KEY WORDS: **Interruption of the aortic arch — Aorticopulmonary septal defect — Aortic arch atresia — Pulmonary artery abnormality**

Interruption of the aortic arch is usually associated with a ventricular septal defect, while aorticopulmonary septal defect (APSD) is commonly associated with an intact ventricular septum and normal aortic arch [21, 23]. Deviation from these usual patterns occurs in a developmental complex characterized by (1) interruption of the aortic arch, (2) APSD, and (3) intact ventricular septum. Isolated case reports of interruption of the aortic arch in association with APSD are present in the literature, and additionally many reports devoted to general reviews of interruption of the aortic arch may include one or more such cases.

Because interruption of the aortic arch in combination with APSD is a correctable condition, it is important to define anatomic details and variations of this complex. The purpose of this communication is to present a review of the anatomic details of 15 cases (United Hospitals of St. Paul and the University of Minnesota, five cases; The Hospital for Sick Children, Toronto, seven cases; The New England Regional Infant Cardiac Program Registry, anatomic details available in two of the three registry cases; St. Joseph's Hospital, Phoenix, Ariz., one case) collected from several institutions with which one or another of the authors is associated and of 31 additional cases found in the literature.

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Definition of Terms

Described in the literature are various types of interruption of the aortic arch and APSD. Since in this review the two anomalies mentioned were each subdivided, where possible, according to accepted subdivision, it is appropriate to summarize the characteristics of the types of interruptions of the aortic arch and the types of APSD.

Interruption of the Aortic Arch

In 1959, Celoria and Patton [9] reviewed the 28 known cases of interruption of the aortic arch and classified them according to the site of the interruption as belonging to one of three types: A, B, or C. Type A interruption of the aortic arch lies distal to the left subclavian artery and is due to a "regression or atrophy of the segment of the aortic arch between the ductus arteriosus and the left subclavian artery." If the interruption of the aortic arch occurs between the left carotid and left subclavian arteries, it is type B and "appears to represent a failure of formation of the fourth left arch." The third type of interruption of the aortic arch, type C, occurs between the innominate and left carotid arteries and, according to the authors, may be "a partial or complete failure of formation of the left third and fourth arches and persistence of the dorsal aorta between these arches as the left common carotid artery, or failure of connection of the outgrowth from the aortic sac with the third and fourth arches and fusion of these two arches to form the left common carotid artery."

Aorticopulmonary Septal Defect

Both Richardson [26] and Mori [22] and their respective associates have recognized that APSD may occur at various sites within the pulmonary arterial tree. Separate and different classification schemes have been proposed by each of these authors to categorize APSD into types.

By the classification of Richardson et al, an APSD may exist as one of three types: I, II, or III. Type I APSD of Richardson et al is "located on the posteromedial wall of the ascending aorta just above the sinus of Valsalva," while defects that lie more distally and "involve the origin of the right pulmonary artery from the main pulmonary artery" are classified as type II defects. Type III APSD of Richardson et al consists of origin of the right pulmonary artery from the ascending aorta, while there is no defect in the left or main pulmonary arteries.

On the other hand, Mori et al do not include origin of the right pulmonary artery from the ascending aorta as one of their three types of APSD. In the classification scheme of Mori et al, both type I and type II APSD remain as described by Richardson et al. Type III APSD of Mori et al (combined or total defect), however, involves the entire length of the pulmonary trunk from immediately above both semilunar valves to the level of the pulmonary bifurcation and the proximal part of the right pulmonary artery.

Since our specimens conform more closely to the three types of APSD described by Mori et al, we have employed their classification scheme to describe them.

Results

This section will consider the following subjects: type of interruption of the aortic arch, the nature of the APSD, and the observed intracardiac and extracardiac associated malformations found in our 15 cases and in the 31 cases found in the literature with interruption of the aortic arch and APSD.

Type of Aortic Arch Interruption

Information as to the type of interruption of the aortic arch was available in 37 cases (Table). In 30 cases the interruption was distal to the left subclavian artery (type A). There was a normal brachiocephalic branching pattern in 28 of these 30 cases, in that there arose successively from the arch the innominate, the left carotid, and the left subclavian arteries, and the descending aorta lay to the left of the spine (Fig. 1**a**).

The right subclavian artery was aberrant, arising from the desending aorta in one of these 30 cases (our case 1) (Fig. 1**b**). Finally, in one of the 30 cases with type A interruption the descending aorta was right-sided [12] (Fig. 1**c**). In this case the interruption of the arch was clearly beyond all of the branches of the arch, but the position of the ductus, whether right- or left-sided, was not stated. While diagrams and photographs present the branching of the arch as in the normal manner, one is led to consider that the case may represent basic right aortic arch and that the true branching was a mirror image of that illustrated in the quoted reference.

In seven of the original 37 cases the site of interruption lay between the left common carotid and the left subclavian arteries (type B interruption) (Fig. 1**d**). Otherwise, the brachiocephalic branching pattern was normal. There were no cases of interruption of the aortic arch proximal to the left carotid artery (type C interruption) identified.

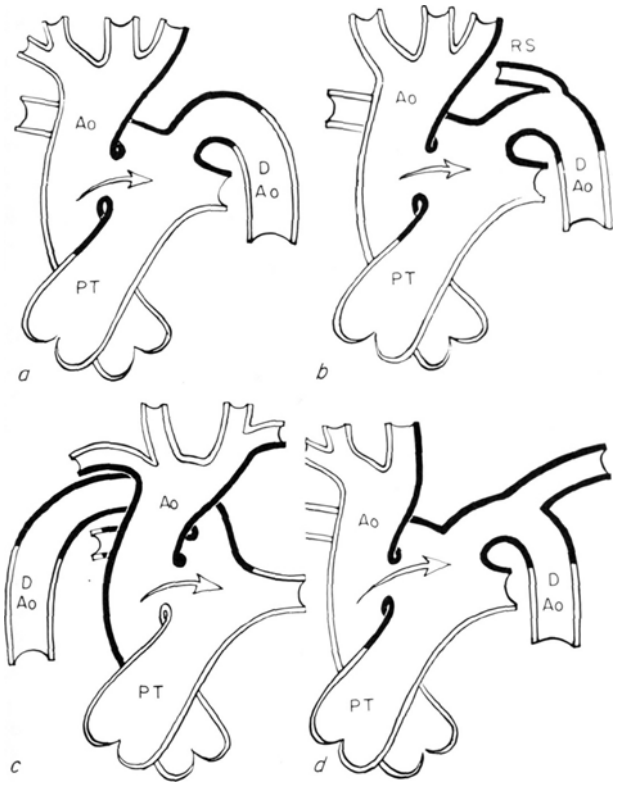

Fig. 1. Anatomic variations of the aortic arch seen in interruption of the aortic arch associated with aorticopulmonary septal defect (APSD) (37 cases). (For simplicity, APSD is illustrated schematically, without regard to type.) **a** Type A interruption of the aortic arch with normal brachiocephalic branching pattern (28 cases). Usual branches arise from aortic arch. **b** Type A interruption of the aortic arch with aberrant right subclavian artery (one case). The right subclavian artery arises anomalously from the descending aorta. **c** Type A interruption of the aortic arch with right-sided descending aorta (one case). The brachiocephalic branching pattern is a mirror image of that illustrated in Fig. 1a. **d** Type B interruption of the aortic arch with normal brachiocephalic branching pattern (seven cases). The left subclavian artery arises from the descending aorta. *Ao*, ascending aorta; *D Ao*, descending aorta; *RS*, right subclavian artery; *PT*, pulmonary trunk.

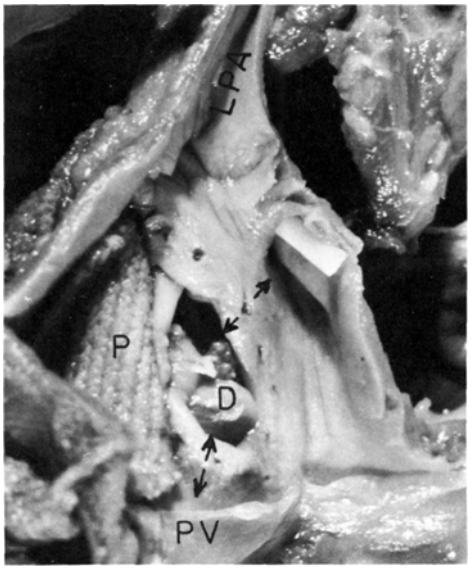

Fig. 2. Type I aorticopulmonary septal defect (APSD) in a case with associated interruption of the aortic arch. Interior of pulmonary trunk, left pulmonary artery (*LPA*) and opened pulmonary valve (*PV*). The posterior line of sutures used to secure the patch (*P*) closing the APSD has been removed and the patch has been reflected anteriorly and toward the specimen's right. The APSD (*D*) is thereby exposed. An aorticopulmonary septum separates the APSD proximally from the pulmonary valve (*arrows*), while a second portion of this septal tissue separates the defect distally (*arrows*) from the origin of the right pulmonary artery (*probe*).

Type of APSD

In 31 of the 46 cases in this study, the type of APSD was either known or adequately described. In 15 others (14 from the literature in which no further anatomic data were available and one from our group in whom only catheterization data were available), the reports contained inadequate information from which conclusions could be drawn as to the type of APSD. The APSD in 11 cases was found to be type I (proximal defect of Mori et al). In each of these cases a partition or septum separated the proximal portion of the ascending aorta from the proximal pulmonary trunk, and the anteroinferior border of the APSD arose distal to this septum. Similarly, the distal edge of the APSD was separated from the origin of the right pulmonary artery by an aorticopulmonary septum (Fig. 2). The APSD was type II or "distal" in 15 cases. In these cases there was no tissue present between the distal edge of the APSD and the origin of the right pulmonary artery, although the proximal septum remained intact. In several cases the right pulmonary artery appeared to arise partly from the posterior surface of the ascending aorta (Figs. 3**a** and **b**). The APSD was "total," conforming to the type III defect of Mori et al in five cases. In these cases, while aortic and pulmonary valves were separated by a broad anular ridge, no thin partition or septum was present beyond the anular ridge, and the APSD extended distally to include the origin of the right pulmonary artery (Fig. 4).

Status of Ventricular Septum

The status of the ventricular septum was known in 38 cases (Table). In 32 of these the septum was intact, while in six cases a ventricular septal defect

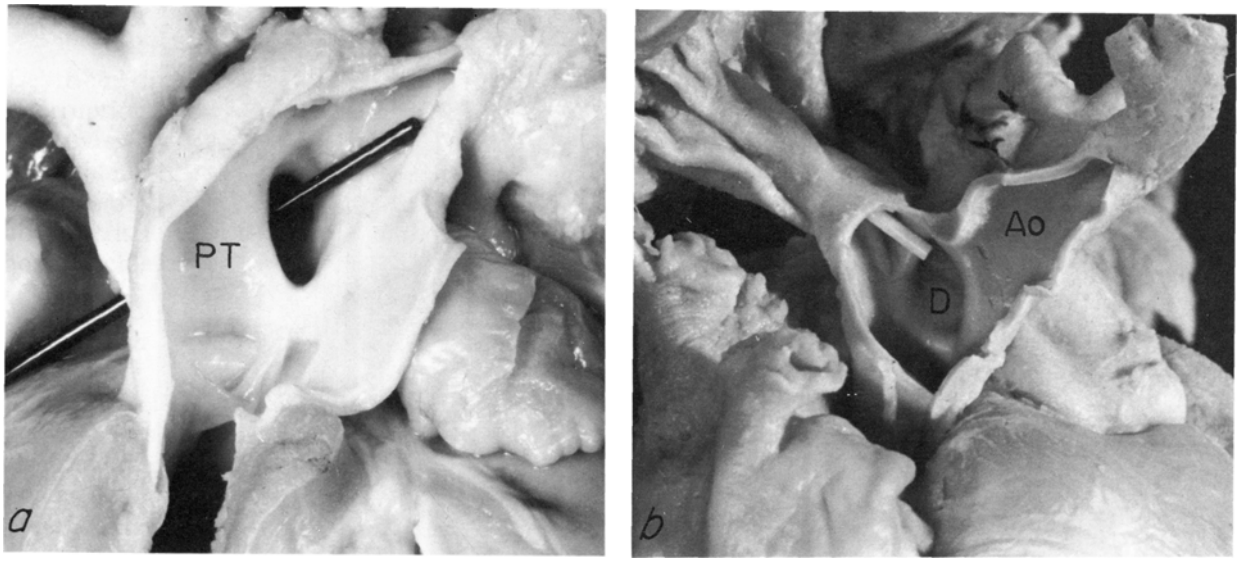

Fig. 3. Type II aorticopulmonary septal defect (APSD) in a case with associated interruption of the aortic arch. **a** Pulmonary trunk (*PT*) with probe in APSD. Aorticopulmonary septal tissue separates the pulmonary valve from the proximal edge of the defect, while the distal extent of the defect includes the origin of the right pulmonary artery. **b** Interior of ascending aorta (*Ao*). The right pulmonary artery (*probe*) arises from the aorta. The APSD (*D*) lies near the origin of the right pulmonary artery.

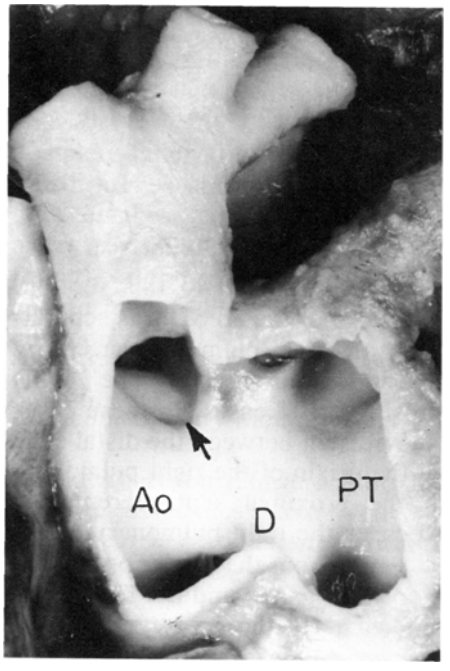

Fig. 4. Type III aorticopulmonary septal defect (APSD) in a case with associated interruption of the aortic arch. The anterior walls of the ascending aorta (*Ao*) and of the pulmonary trunk (*PT*) have been removed to expose the interior and the APSD (*D*). A separate semilunar valve was present at the base of each great vessel. The proximal edge of the APSD lies directly above the valve anuli, and no aorticopulmonary septum is present. The APSD extends distally to include the origin of the right pulmonary artery (*arrow*).

was present (Table). The ventricular septal defect was "isolated" in five of these cases, while in the sixth case the ventricular septal defect was subpulmonary in nature and occurred as part of a complex lesion, including aortic atresia [27].

Both the site of the aortic arch interruption and the status of the ventricular septum were known in 32 cases. In these cases a ventricular septal defect was more likely to occur with a type B interruption of the aorta (three of seven cases) than with a type A interruption of the aorta (three of 25 cases) (Table).

Other major intracardiac defects, such as transposition of the great arteries or origin of both great arteries from the right ventricle, both of which occur in interruption of the aortic arch with some frequency [21], did not occur in our cases.

The report of Becu et al in 1955 [2] emphasized that in interruption of the aortic arch, the usually occurring ventricular septal defect lies partly in a subpulmonary position and that beyond the defect a spur of tissue gives rise to subaortic obstruction. The presence of subaortic obstruction in interruption of the aortic arch associated with APSD was difficult to determine in those cases identified by literature search because little attention has previously been given to it [13]. In this study then, subaortic obstruction was deemed *absent* if (1) it was not observed by our own examination of the heart at postmortem examination (our cases 3 to 5, 7, 10 to 15); (2) catheterization data demonstrated either no gradient or a gradient of 20 mm Hg or less between the left ventricle and the ascending aorta

Table. Anatomic details in interruption of the aortic arch with aorticopulmonary septal defect

Site of interrupted aortic arch	Status of ventricular septum	Type of APSD I	II	III	Unknown
A	Intact, 22 cases	6 cases: present study, cases 5, 9, 11, 14, 15*; ref. 28	7 cases: present study, cases 1, 4, 6–8; ref. 24, 32	3 cases: present study, case 3; ref. 11, 1	6 cases: ref. 12, 15, 6, 10, 8, 5
	Defect, 3 cases	1 case: present study, case 10	. . .	. . .	2 cases: ref. 14, 19
	Unknown, 5 cases	. . .	3 cases: ref. 29	. . .	2 cases: cited in ref. 15
Subtotal	*30 cases*	*7*	*10*	*3*	*10*
B	Intact, 4 cases	2 cases: present study, cases 12*, 13*; *described also in ref. 20, 13	. . .	. . .	2 cases: ref. 3, 33
	Defect, 3 cases	1 case: ref. 27	. . .	. . .	2 cases: present study, case 2; ref. 31
Subtotal	*7 cases*	*3*	. . .	. . .	*4*
Unknown	Intact, 6 cases	1 case: ref. 18	5 cases: ref. 4	. . .	. . .
	Unknown, 3 cases	. . .	. . .	2 cases: cited in ref. 11, 30	1 case: ref. 25
Subtotal	*9 cases*	*1*	*5*	*2*	*1*
Grand total	*46 cases*	*11*	*15*	*5*	*15*

*described in ref. 16

(our cases 6 and 9 [11, 12, 14]); or (3) description of postmortem examination specifically stated that subaortic stenosis was absent [32]. By these criteria, the classic picture of subaortic stenosis was not present in any of the 16 adequately documented cases.

Miscellaneous cardiac malformations consisted of atrial communications in nine cases: secundum atrial septal defect, four cases (our cases 2, 5, 14 [19]); patent foramen ovale, five cases [3, 24, 27, 28, 32]. A persistent left superior vena cava was present in two cases (our case 5 [15]). In one case the left coronary artery appeared to arise anomalously from the pulmonary side of the APSD [8]. In one case a right pulmonary vein returned to the right atrium [24]. The case of aortic atresia and ventricular septal defect has been mentioned in an earlier section.

Extracardiac Abnormalities

In five cases major whole-body abnormalities were noted: Down's syndrome [31], Vater's association (our case 8), probable Di George syndrome (our case 14), probable fetal alcohol syndrome [28], and polydactyly of the right hand [24].

In the 30 cases where the sex was stated there was a predominance of males (18 cases) to females (12 cases).

Birth weight was available in 18 of the 43 cases, and the average weight of these infants was less than normal. One child, born at 34 weeks' gestation, weighed 1.64 kg [28]. If he is excluded, the average birth weight of the remaining 15 infants is 2.88 kg, ranging from 2.3 to 4.1 kg.

Discussion

Interruption of the aortic arch is a rare congenital anomaly. The condition was found in 24 of the 2,251 infants entered into the New England Regional Infant Cardiac Program registry (NERICP) between 1969 and 1974 [25]. Of these 24 infants, three had an associated APSD. Similarly, the incidence of APSD by itself is also low, occurring only seven times (0.3%) in the NERICP series. Of these seven cases with APSD, however, three had an associated interruption of the aortic arch (42.8%), and, of interest, in one of the remaining four of these cases there was an associated coarctation of the aorta [25].

The occurrence of various cardiac anomalies with APSD has been analyzed by others [7, 23]. Blieden and Moller [7] reported two cases of interruption of the aortic arch among their 17 cases of APSD. On the other hand, Neufeld et al [23] found no cases of interruption of the arch and only three cases of coarctation of the aorta among the 66 cases of

APSD in their review. The discrepancy between these results and those of the NERICP can be partially explained by examining the patient populations involved in these studies. All cases in the NERICP were of patients younger than 1 year when entered into the files. It would be reasonable to assume that nearly all patients with interruption of the aorta and APSD will present in infancy and thus be found by a registry such as the NERICP. On the other hand, some cases of isolated APSD that present in later life might not be found by such a registry. Thus, it is not unexpected that the reviews of Blieden and Moller and of Neufeld et al, both of which contain cases of patients ranging in age from less than 1 month to well into adulthood (40 years), contain a smaller percentage of cases of APSD and interruption of the aortic arch.

Thus, the recognition by Berry et al [4] of a specific syndrome consisting of APSD, aortic origin of the right pulmonary artery (type II APSD), and interruption of the aortic arch appears valid. Our findings indicate, however, that any type of APSD, not only the type II defect noted by Berry et al, may be associated with interruption of the aortic arch. Interestingly, a single case of interruption of the aortic arch has been reported with no APSD but with aortic origin of the right pulmonary artery [17] (type III APSD of Richardson et al [26]).

In contrast to the classic interruption of the aortic arch with ventricular septal defect in which subaortic stenosis may be associated, the latter condition has not been noted either clinically or at postmortem examination when APSD is associated with interruption of the aortic arch. Thus, the theoretical argument given for interruption of the aorta in these cases, namely, obstruction of the left ventricular outflow tract and diversion of the fetal circulation across the ventricular septal defect into the pulmonary circulation, may not always be the same mechanism that leads to interruption of the aortic arch when it is associated with APSD. The diversion of aortic arch blood into the pulmonary circulation via APSD, as proposed by Berry et al ("aortic arch steal"), may be a plausible explanation for this combination of defects [4]. The case of Rosenquist et al [27], consisting of aortic atresia, interruption of the aortic arch, APSD, and subpulmonary ventricular septal defect, is, however, unique in that unusual and extreme left ventricular outflow obstruction, as well as an APSD, coexist in the same specimen.

Within the group of cases of interruption of the aortic arch and APSD reviewed by us, type A interruption predominated. This is in contrast to the nearly equal occurrence of types A and B interruption when they are reported without regard to intracardiac defects [21].

Additionally, we found that although the ventricular septum is usually intact, it is not always so. From our series, a ventricular septal defect was more common with a type B interruption of the aortic arch and APSD than when the interruption was of the type A variety.

Although the site of the APSD was variable, we were unable to correlate the APSD with either the type of aortic arch interruption or the presence of ventricular septal defect.

Finally, we noted that infants with the condition appear to be small for gestational age. This may perhaps complicate the surgical repair as in our series where two of our operated cases (cases 5 and 7) showed that surgical closure of the APSD (type II) was complicated by inadvertant occlusion of the origin of the right pulmonary artery.

Conclusions

In our review of 46 cases of interruption of the aortic arch and associated APSD we found that all but one of the cases reviewed showed potentially correctable lesions. We have condensed the preceding data into "rules of thumb" for the preoperative evaluation of any patient in whom interruption of the aortic arch or APSD are present or suspected.

1. In infantile cases of interruption of the aortic arch in whom the ventricular septum is intact, look for the APSD that almost certainly exists.
2. In infantile cases of APSD, establish the integrity of the aortic arch and the normal origin of both coronary arteries.
3. In cases of interruption of the aortic arch and associated APSD, define the situs of the descending aorta (right- or left-sided).
4. In cases of interruption of the aortic arch and APSD, define the type of the interruption of the aortic arch (A, B, or C), and if the type of interruption is type B, look for an additional left-to-right shunt at the ventricular level.

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Addendum. Since submission of this article, we have examined one further case of type A interruption of the aortic arch, intact ventricular septum, and type I APSD with associated secundum atrial septal defect in a newborn pre-term male infant whose additional extracardiac defects included tracheoesophageal fistula, bronchus suis and single umbilical artery.