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Case Report

Aortopulmonary Septal Defect

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Summary. The heart of a 42 year old man with a total defect of the aorto-pulmonary septum is described. Such cases should not be called partial truncus arteriosus persistens, since they have a normal bulbus whereas in cases of t.a.p. it is defective.

Aortopulmonary defects differ from the truncus arteriosus persistens (t.a.p.) by having a normally formed bulbar septum, yet the partition between the aorta and pulmonary arteries is defective distal to the semilunar valves. The defect between these two arteries may vary from a small opening of only a few millimeters to one involving the entire septum (Hudson, 1965). The symptoms of the latter can easily simulate those of the t.a.p. Just as the ventricular septal defects are not called partial common ventricles, the aortopulmonary fenestrations should not be called partial t.a.p. (Van Pragh and Van Pragh, 1965). The complete defect of the septum trunci is, in the opinion of several authors, a special form of the t.a.p. (Collet and Edwards, 1949). Van Pragh and Van Pragh (1965) and Keith *et al.* (1967) even refer to the complete absence of the truncal septum as the real true truncus arteriosus persistens, since in their opinion the classic t.a.p. is not only caused by deficient septation of the truncus but also by extreme underdevelopment of the pulmonary infundibulum associated with the bulbar septal defect. Thus these authors point out that the term truncus arteriosus persistens inadequately describes the changes of the classic t.a.p. Not only the common truncus, but the malformation of the distal bulbus, the ventricular defect and the abnormal development of the semilunar valves are all part of t.a.p. Goerttler (1969) even suggests that the true cause of the t.a.p. is the deficient formation of the distal bulbus.

We believe therefore a defect of the truncal septum associated with a regularly formed distal bulbus should clearly be distinguished from the t.a.p.

Clinical and Pathological Data

Sixteen years after the angiographic diagnosis of an aortopulmonary defect had been made and a surgical attempt to repair his defect failed because of its large size, this 42 year old man died of congestive heart failure.

At autopsy (SN 401/71, Path. Inst. Univ. Heidelberg) the heart weighed 670 gm. The apex was rounded. No outer abnormalities were visible. Right atrium regularly developed. Foramen ovale closed. Cusps and chordae tendineae of the tricuspid valve, delicate, circumference

Fig. 1. Ventral view into the right ventricle. Above the pulmonary cusps the common aorto-pulmonary trunk may be seen separated by a fold between pulmonary and aortic ostium

12 cm. Right ventricle dilated, myocardium hypertrophied, pale-red and flabby. At the base it was 12 mm thick. Trabecular and papillary muscles hypertrophied. Outflow tract and conus pulmonalis widely patent. Pulmonary valves normally developed with three delicate semilunar cusps. Circumference 9 cm. Left atrium moderately dilated. Mitral valve and chordae tendineae delicate and transparent. Circumference 10.5 cm. Left ventricle dilated, with enlarged infra-papillary space. Membranous portion of the septum interventriculorum triangular, of normal size (ca. 0.8 cm^2). Myocardium flabby, pale-red and about 1.3 cm thick. Outflow tract and aortic ostium normal. Three delicate, regular semilunar cusps. Coronary ostia normally situated in the sinus of Valsalva. On opening the right ventricular outflow tract and on cutting through the pulmonary valve a large vascular sac was exposed. It extended from the upper level of the aortic and pulmonary orifices upwards to the aortic arch. Between the aortic and the pulmonary ostia a $10 \times 15 \times 2$–3 mm crescent-like fold arose behind the right pulmonary cusp and branched at the junction between the right and posterior semilunar cusp, forming two folds (5 cm long and 2 cm broad) that extended spindle-like to the region of the left carotid artery. In the mid region of this sac-like aorto-pulmonary vessel the pulmonary arteries branched off normally into the lungs (Figs. 1 and 2). The aorta arched to the left and gave off its branches in a normal manner. The common vascular trunk was coated by an intima

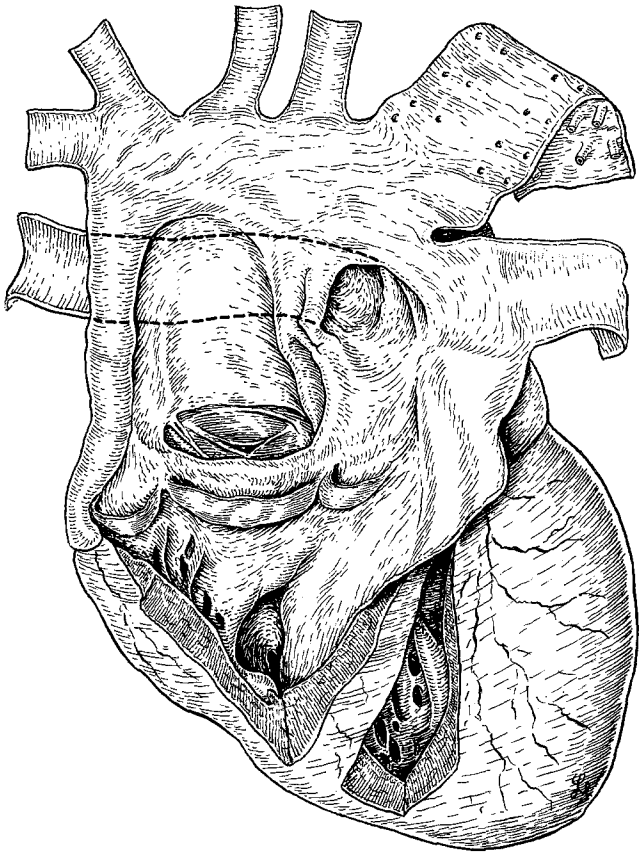

Fig. 2. Sketch of ventro-cranial view

with multiple arteriosclerotic plaques. Microscopically the fold consisted of a dense, partially hyalinized collagenous tissue without muscle fibers. The other organs did not show significant pathological changes.

Discussion

The directions of flow of the intracardial bloodstreams and with them the positions of the endocardial septa are determined by the form of the myo-epicardial coat and the hemodynamic factors it modifies (Goerttler, 1969). Spitzer (1923) first stressed the importance of hemodynamic factors for the development of the cardiac septae. Doerr (1952, 1953) referrred to the complex movements of the ventricular outflow tract as bulbus-torsion. Septations in this region, whether regular or abnormal, depend on this bulbus torsion.

The classic t.a.p. is always associated with a defect in the distal bulbus since it represents a defect in the bulbar septum and seldom also a shortening of the entire bulbus (Van Pragh and Van Pragh, 1965). The common aorto-pulmonary vessel and the malformed septum might be the result of a deficient formation and torsion of the bulbus, so that the beds of the future pulmonary and aortic

bloodstreams are parallel. Two parallel flows do not overcross and are unable to produce-pressure-free zones needed to form septa (Goerttler, 1955). This hypothesis might explain how the aortopulmonary septal defect in t.a.p. develops. Another explanation might be a large defect of the distal bulbar septum resulting in a faulty arrangement of the bloodstreams and would agree with the concepts of Abbott (1927), Humphreys (1931), Lev, Saphir (1935), Collet and Edwards (1949), De la Cruz and Rocha (1955) and Nadas (1963), who consider the cause of the t.a.p. to be a deficient formation of the bulbar ridges. We believe however that a deficient septation can only be a secondary phenomenon since the endocardial bulges and ridges responsible for the formation of the septa show only minimal proliferation compared with the very actively proliferating myoepicardial coat, which consequently is more likely to be damaged by certain noxious agents (Goerttler, 1956; Grohmann, 1961). If the formation of the bulbus is disturbed causing abnormal blood flow that leads to a deficient development of the septa, it seems acceptable to postulate, that when the bulbus is normally formed with regularly arranged bloodstreams, septation should develop normally. Hence, single, complete defects of the aorto-pulmonary septum should not exist if the bulbus were well-formed. However, two other cases have been described, in which the septum showed an identical defect. (Bain and Parkinson, 1942; Van Pragh and Van Pragh, 1965, with an illustration from Duckworth). In their two cases and in our case the bulbar regions and arterial orifices were normally formed with normal semilunar valves. The bloodstreams after the passage through the orifices should have taken the right way, overcrossing each other, and should have hemodynamically led to the formation of the truncal septum. If one conceives an imaginary septum at the above described fold separating the two vascular beds, the truncal and the bulbar regions seem to be normally formed and are easily recognised when examined from the common trunk, aorta and pulmonary arteries. The cause of this rare malformation of the septum cannot be a disturbed development of the bulbus. On the contrary its cause is to be regarded as a malformation of the truncus.

The boundary between bulbus und truncus according to most investigators is seen at the level of the semilunar valves (Tandler, 1912; Pernkopf and Wirtinger, 1935; Kramer, 1942; De Vries and Saunders, 1962; Asami, 1969), although De la Cruz and Rocha (1955) regard that boundary as arbitrary. Kramer (1942) however relates the semilunar valves to the truncal ridges, whereas Asami (1969) believes the latter forms as the result of the transformation of the distal bulbar ridges. The presence of musculature in the aortic bulb also strongly suggests this region belongs to the distal bulbus (Asami, 1969), whereas the truncal septum consists of a cell-rich tissue with large nuclei, resembling the noncardiac splanchnic mesoderm that forms the aorta and pulmonary artery (Tandler, 1912; De Vries and Saunders, 1962). Even if we do not believe that the formation of the septum occurs independent of the blood streams and is unimportant for their direction of flow it seems that special regions of the truncal septa have a certain histogenetic specificity that enables them to develop in a special way. Since in our case the bulbar region and semilunar valves were normally developed, the primary cause of the septal defect should not be looked for intracardially but rather extracardially in the non-cardiac splanchnic mesoderm. Hence it appears that the

isolated defects of the truncal septum have nothing in common with the true t.a.p. except the large common vessel. They represent rather malformations that are not dependent on the development of the bulbar ridges and the bulbar torsion. Therefore these three cases should not be classified as type 5 of the t.a.p. (Collet and Edwards, 1949) or as type B in the classification by Van Pragh and Van Pragh (1965). Edwards (1960) questions the existence of these malformations and tries to imply a different mechanism for the defect of the truncal septa. He mentions a case with a simultaneous rupture of the wall of the ductus arteriosus, aorta and pulmonary artery, resulting in the formation of a "common trunk". Such a defect, none-the-less, has only a structural similarity to the discussed problems and does not represent a true malformation. In our case and in the others cited we were unable to find any evidence that the defect was caused by such a rupture. After analysing these cases we do not want to draw any conclusions about an independent development of the truncal septum as postulated by Pernkopf and Wirtinger (1935). As our case indicates, however, there may be a certain degree of independency in the development of single "meral" cardiac regions, particularly during the formation of a defect (Doerr, 1943). Our case proves that the septation is not the critical event in the partition of the bulbar-truncal region, but rather it is the bulbar torsion as conceived by Doerr (1943, 1950, 1952, 1955), since as postulated by Doerr and as shown by this case, a regular formation of the truncal region is possible even without a septum.

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