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 trunci arteriosus persistens, since they have a normal bulbus whereas in cases of t.a.p. it is defective.

Aortopulmonary defects differ from the trunci 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 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 trunci septum as the real true trunci arteriosus persistens, since in their opinion the classic t.a.p. is not only caused by deficient septation of the trunci but also by extreme underdevelopment of the pulmonary infundibulum associated with the bulbar septal defect. Thus these authors point out that the term trunci arteriosus persistens inadequately describes the changes of the classic t.a.p. Not only the common trunci, 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 trunci 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
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 infrapapillary space. Membranous portion of the septum interventriculorum triangular, of normal size (ca. 0.8 cm³). 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 × 15 × 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. 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.