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On the Formal Genesis of the Eisenmenger Complex

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Summary. The arrangement of the musculature at the aortal conus is eminently important for the interpretation of a riding aorta in the Eisenmenger complex. In the case discussed, a muscular bar coursing into the right ventricle to the crista supraventricularis was present between the annulus aortae and the VSD despite a riding aorta. This anomalous muscular connexion between the conus aortae and the right ventricle owes its existence to a *primary heterotopia* of the aorta caused by a premature arrest of the *vectoral bulbus torsion.* The subannular muscle tissue was of bulbometampullar origin.

Zusammenfassung. Zur Deutung der reitenden Aorta bei dem Eisenmenger-Komplex ist die Anordnung der Muskulatur am aortalen Conus von besonderer Bedeutung. Beim erörterten Falle liegt zwischen dem Annulus aortae und dem Ventrikelseptumdefekt trotz des Überreitens der Aorta ein Muskelbalken, der sich in die rechte Kammer bis zur Crista supraventricularis fortsetzt. Dieser anomale muskuläre Zusammenhang zwischen dem aortalen Conus und dem rechten Ventrikel läßt auf eine *primäre Heterotopie* der Aorta durch einen Arrest der *vektoriellen Bulbusdrehung* schließen. Das subannuläre Muskelgewebe stellt Muskulatur bulbometampullärer Herkunft dar.

The indisputable finding of the aorta riding over a ventricular septal defect in the tetralogy of Fallot and Eisenmenger complex can be interpreted in the following two ways: According to Doerr (1951, 1952, 1955), the riding aorta is the result of an arrest of the vectoral torsion of the bulbus. Bankl (1971 a, b) attributes it to an absence of muscle tissue under the annulus. It has already been demonstrated that in the tetralogy of Fallot, the anatomical relationships of the annulus aortae to the neighboring structures depend upon a primary heterotopia of the aorta in the sense of the Doerr concept (Chuaqui, 1971). In this report, we wish to illustrate that, as in the case of an Eisenmenger complex, relationships similar to a riding aorta, especially an anomalous muscle attachment between the annulus aortae and the right ventricle, could exist.

Case Report. A 5 year old girl, the first child of a 23 year old healthy woman. Uncomplicated pregnancy and delivery. Postpartum apnea which responded immediately to O_2 therapy. Further development was generally retarded marked with a slight decrease in vitality. Congenital "heart disease" recognized at birth. The diagnosis of VSD with Eisenmenger reaction (shunt reversal: crosshunt with 45–50% left-to-right shunt and 20% right-to-left shunt) and pulmonary hypertension in November 1971. Operation in February 1972. Closure of the VSD with a prosthetic patch and suturing over of a large, wide patent foramen ovale. Postoperative cardiac decompensation, cardiogenic shock.

Pathologic-Anatomical Finding. A large, overweight heart (190 g). Specimen fixed in formalin, right ventricle wallthickness measured 10,5 mm, the left 9 mm. Both ventricles, especially the right, demonstrate a pronounced trabecular relief. The papillary muscles appear very large. No cardiac or pulmonary vein anomalies. In the area of the fossa ovalis is found an 18 mm long and 3 mm wide ASD surgically closed to a slit. Mitral ostium (7.2 cm dia-

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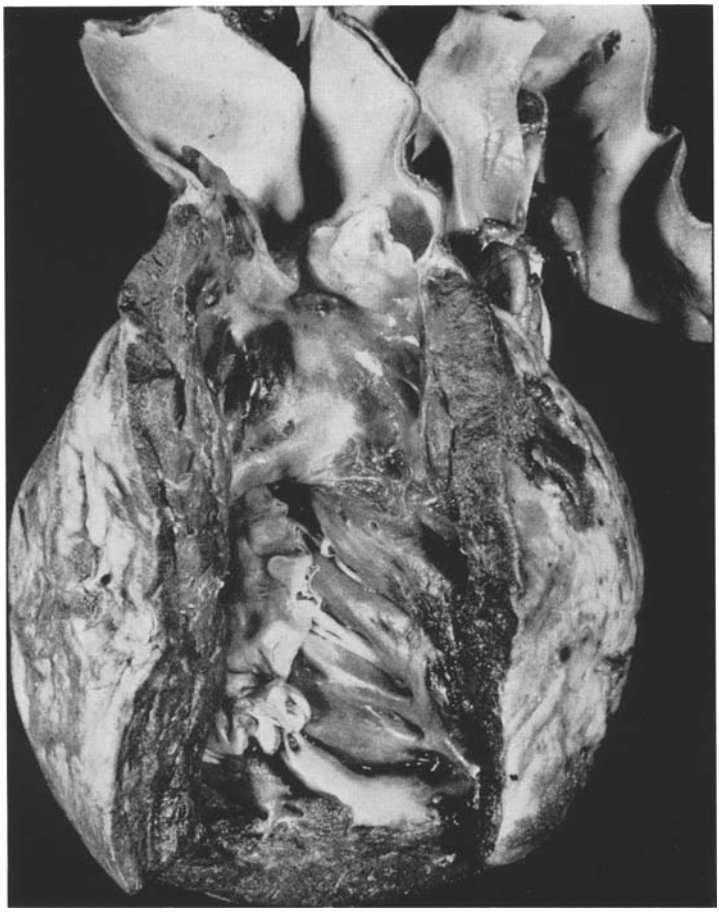
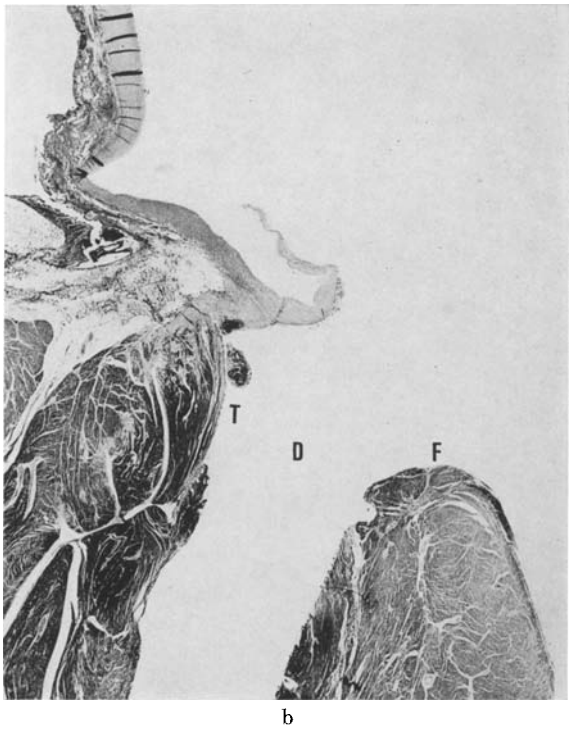


Fig. 1. View into the markedly hypertrophied right ventricle. The interventricular septum defect appears directly beneath the crista supraventricularis. No pulmonary artery stenosis. Dilatation of the pulmonary trunk. (Autopsy number: 167/72, Pathology Institute, Univ. of Heidelberg)

Fig. 2. a VSD as seen from the left. Inferior border of the defect is the interventricular septum ridge. Beneath the annulus aortae is a muscular bar which courses behind the interventricular septum towards the right ventricle. Part of the tricuspid valve is seen near the defective pars membranacea. b Frontal section through the interventricular septum at the level of the defect. *F* interventricular septum ridge; *D* defect; *T* muscular trabecula under the annulus aortae. Magnification ×6; H'E stain (autopsy number 167/72, Pathology Institute, Univ. of Heidelberg)

a



b

Fig. 2a and b

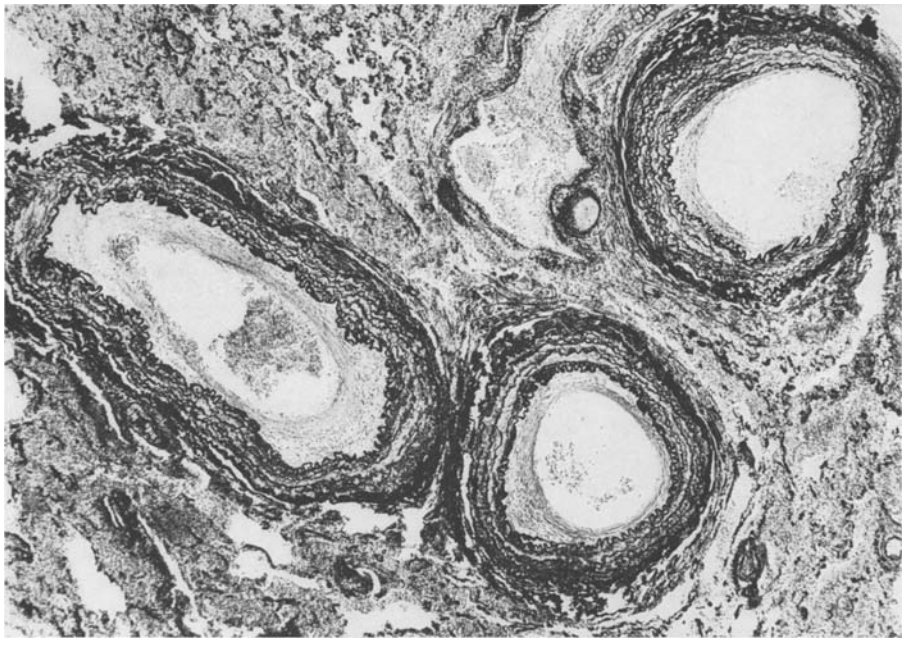


Fig. 3. Medium-sized pulmonary artery branches. Note the thickening of the media and the intima hyperplasia (Pulmonary artery sclerosis Grade III). Magnification circa ×40; van Gieson-Verhoeff stain (autopsy number 167/72, Pathology Institute, Univ. of Heidelberg)

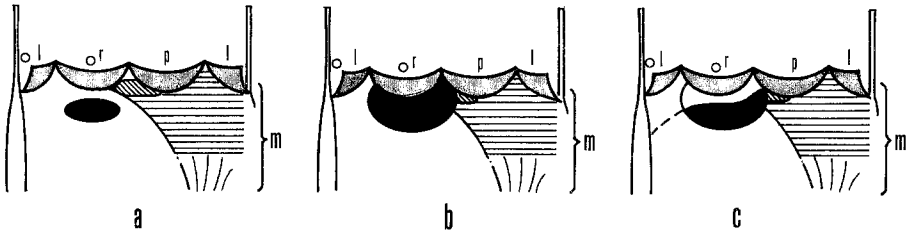


Fig. 4a—c. Schematic drawing of the anatomical relationships of VSD situated in the posterior portion of the anterior interventricular septum. View from the left: *m* septal cusp of the mitral valve; oblique shading, the pars membranacea; *r* right, *p* posterior and *l* left semilunar cusps of the aorta. a Defect in zone B II fully circumscribed by musculature. b Defect in zone B II and the pars membranacea (zone B III) without a muscular border under the annulus aortae. c Topography of the defect as in b. The muscular trabecula sits beneath the annulus aortae (counter ridge). Dashed line marks its course behind the interventricular septum to the crista supraventricularis

meter) and tricuspid ostium (8.2 cm diameter) unremarkable. The pulmonary ostium (4.3 cm diameter) possessed normal dimensions but had only two semilunar cusps of unequal size. The larger of the two demonstrates a rudimentary commissure at its base. Dilated pulmonary artery trunk with thickened intima (Fig. 1). The aorta, measuring 4.5 cm in diameter at its base, rode on the interventricular septum in which a high defect was present. The defect measured 1.3 cm in diameter and approximated a rectangle. On the left side of the interventricular septum, the defect reaches from the right half of the posterior cusp to the left

half of the right cusp of the aorta. On the right side of the interventricular septum, the defect was situated inferior and posterior to the crista supraventricularis and is insufficiently closed by a prosthetic patch. The caudal rim of the septum defect is formed by the interventricular septum ridge. Cranially, the defect borders on a 4 mm muscle bar which simultaneously supports the cusps of the riding aorta. This bar comes to view under the commissure between the right and posterior cusps, forming an acute angle between the right ridge of the posterior cusp and the insertion of the medial mitral cusp (Fig. 2a). The bar courses 5 mm behind the interventricular septum ridge toward the right, enters the posterior surface of the crista supraventricularis, meshing with it between the pars septalis cristae. Microscopic examination of the VSD region demonstrates the trabecula to be constructed of muscle tissue. The left bundle branch transverses over the ventricular septum ridge inferior to the defect (Fig. 2b). The histological examination of the lungs yielded pulmonary artery sclerosis Grade III (Heath and Edwards, 1958; Fig. 3).

Summary of Pathologic-Anatomical Diagnosis. Riding aorta; High VSD in the region of the pars membranacea of the interventricular septum; pulmonary artery sclerosis; right heart hypertrophy (socalled Eisenmenger syndrome); slit-like ostium secundum defect of the atrial septum; postoperative status for VSD and secundum defect repair; cardiac decompensation in cardiogenic shock.

Discussion

In 1897 Eisenmenger described the pathophysiologic signs in a case of a high-seated VSD in which he also mentioned the accompanying right heart hypertrophy and dilatation of the pulmonary artery. According to the classification of Rokitansky (1785), the ventricular septum defect was situated in the posterior portion of the anterior septum, that is, where most VSDs are located (Becú, 1956; Chuaqui, 1968; Doerr, 1967; Goerttler, 1960). A description of the muscular rim of the defect was not included in the report. Although Eisenmenger did refer to the aorta riding directly over the defect, the position of the great arterial vessels was reported as otherwise normal. A second paper by Eisenmenger in 1898 dealt with the phenomenon of the "riding aorta" in defects in the posterior portion of the anterior interventricular septum. He concluded the ensuing by dissection of fixed normal hearts: Such a septum defect can present without a riding aorta, however never vice-versa. In modern clinical cardiology his original findings of right heart hypertrophy and pulmonary artery wall changes secondary to right ventricular hypertension are called *Eisenmenger reaction* in various congenital heart defects. The Eisenmenger complex refers only to a VSD with pulmonary hypertension (Wood, 1958). The characteristic of the complex — riding aorta — has lost its significance in clinical cardiology (Brammel *et al.*, 1971). However, this phenomenon and its morphology has received renewed interests, especially from Bankl (1971 a, b). Using the examination methods of Eisenmenger (1898), he has been able to conclude the following: The right circumference of the aortal ostium which rides on the defect in the posterior portion of the anterior ventricular septum only then gains a connexion with the right ventricle when the superior margin of the defect is not separated by muscle tissue from the annulus aortae, i.e. the presence or absence of a muscular bar arising from the annulus aortae decides wether or not the aorta rides over the septum defect. The position of such a interventricular septum defect includes the pars membranacea (zone B III of the Bersch classification, 1971) and the fossa subinfundibularis — an area between the pars membranacea and the crista supraventricularis — (zone B II of the Bersch classification, 1971) on the right side of the inter-

ventricular septum. On the left side, zone B II (Bersch, 1971) corresponds to the area inferior to the right aortal cusp (Becú, 1956) and zone B III to the pars membranacea. The presence or absence of a muscular base for the annulus aortae determines 2 types of septal defects at this site (Fig. 4a and b). Type *a* with a muscular base appears in VSDs at B II relatively infrequent (5 of 27 cases in isolated defects at this zone in the interventricular septum, Chuaqui, 1968). Type *b* with absent muscular base has been implicated by Bankl (1971 a, b) as the sole reason for the riding aorta over the VSD. With this generalisation, Bankl negates Doerr's concept that the riding aorta can be caused by an arrest in the bulbus torsion (Doerr, 1952, 1955 a, b, 1960) which has been proven to occur by Asami (1969). The presented case fills the requirements for the Eisenmenger complex clinically and anatomically, demonstrating a wide defect in zone B II and B III of the septum which corresponds to neither of the two types mentioned. A muscular bar is present inferior to the annulus aortae in spite of the riding aorta. This bar does not mesh with the interventricular septum ridge anteriorly but courses along the right surface of the interventricular septum to the crista supraventricularis (type c; Fig. 4c). This trabecula proves to be an anomalous muscular connexion between the musculature of the aortal conus and the right ventricle. The overriding of the aorta does not owe its presence to an absence of basal muscular support of the annulus. The anomalous connexion is more likely due to a primary heterotopia of the aorta and composed of subaortal muscle tissue. Its inability to reach the ridge of the septum is the result of a premature arrest of the aortal ostium at the interventricular septum ridge. The question as to origin of the described trabecula is of particular importance. Contrary to Bankl's view, an overriding aorta appears normally in the XVIII horizon in classification of Streeter (Asami, 1969). At this stage, the right aspect of the aortal conus is formed by the counter ridge *B–O* (see Bersch, 1971, 1972). In horizon XIX a suture line between the counter and main ridge can be seen (Bersch, 1972), which also appears in XX horizon, albeit less clearly (Chuaqui and Bersch, 1972). Immediately inferior to this horizontal suture line, in zone B II, lies the left branch of the AV conduction bundle. In older embryos as well as in fully developed fetus without this suture line, the left branch lies inferior to the right aortal cusp and becomes the true bordering structure between the underlying musculature of the primitive interventricular septum and the overlying subaortal muscle tissue of the counter ridge. The reliability of this markation method has been proven by the interpretation of a case with multiple VSDs (Kreinsen and Bersch, 1972). Applying this principle to our case with VSD-type c (Fig. 4c), it becomes evident that the horizontally coursing subannular part of the described trabecula corresponds to the *counter ridge*. The same applies generally to the muscle tissue which courses between the annulus aortae and the defect in VSD type a (Fig. 4a). The oblique right ventricular portion of the muscular bar coursing anterior to the crista supraventricularis is more difficult to interpret. The following hypotheses may shed some light: The counter ridge—bulboauricular ridge—(Bersch, 1971) courses from the anterior endocardial cushion *O* to the bulbar swelling *B* (also see Asami, 1969; Doerr, 1952, 1955 a and b; Pernkopf and Wirtinger, 1933). The formation of the crista supraventricularis in which the musculature of the metampulla and bulbus are included (Asami, 1969; Pernkopf and Wirtinger, 1933) causes a dis-

appearance of the boundaries between the bulbar ridge and swelling. Although the bulbar swelling does not correspond to any anatomical structure in the fully developed heart, its final location can be traced to the area of the muscle of Luschka (Pernkopf and Wirtinger, 1933). In the tetralogy of Fallot the right ventricular part of the muscular bar courses nearly parallel to the pars parietalis cristae, reaching the lateral ventricular wall, i.e. quite some distance from the accepted location of the bulbar swelling *B*. This bar represents no known structure in the normal heart. In our case, this muscular bar ends in an area which could possibly lie within the bulbar swelling *B*, i.e. immediately behind the region in which the pars parietalis cristae and pars septalis meet. It appears resonable that the entire trabeculae in this case represents a displaced bulboauricular ridge.

The presented case has demonstrated the riding aorta in an Eisenmenger complex can also be caused by a disturbance in the vectoral bulbus torsion. The Eisenmenger complex can thereby be correctly included in the existing tetralogical classification of Doerr (Doerr, 1951, 1952, 1955 a and b, 1960).

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