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Applying a Classification Principle of Ventricular Septal Defects to a Case with Several Defects of the Interventricular Septum

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Summary. We have described a human heart with three ventricular septal defects (Swiss cheese septum, Doerr, 1967). The openings, located in various parts of the septum, were analyzed by descriptive embryological principles (Bersch, 1971). We studied how the left bundle of the conduction system was related to the ventricular septal defects; our results confirmed the value of classifying these defects by embryological principles.

Zusammenfassung. Ein Fall einer Herzmißbildung mit drei Ventrikelseptumdefekten (sog. Swiss Cheese Septum, Doerr, 1967) wird beschrieben, Die verschieden lokalisierten Septumlücken werden anhand eines Einteilungsprinzipes nach rein embryologischen Gesichtspunkten (Bersch, 1971) untersucht. Auf die besondere Bedeutung einer Untersuchung des linken Schenkels des Reizleitungssystems bei Ventrikelseptumdefekten zur Erweiterung dieses Einteilungsprinzips wird hingewiesen.

Clinical Data

The patient was a seven year old girl and first child of a 20 year old woman. Though pregnancy and delivery had allegedly been normal, the child's further development became characterized by a frail constitution and reduced stamina. When four years old a heart murmur was diagnosed. In April 1970 the diagnosis of a ventricular septal defect with pulmonary hypertension was established. In May 1970 a surgical repair of the ventricular septal defect was carried out but the child died shortly thereafter in acute cardiac failure.

Gross Pathological Findings

The enlarged 160 g heart measured 7 cm from apex to base. The venous and arterial orifices and their valves were normally developed. The foramen ovale was closed. After formalin fixation, the wall of the right ventricle measured 0.5 cm thick, that of the left ventricle 1.0 cm. The trabecular muscles of both chambers were stout. The posterior papillary muscle of the left ventricle was well-developed with a broad base. It measured 3 cm long, extending two-thirds the height of the ventricle. Its upper end was 1.5 cm thick. The chordae tendineae of the mitral valve were normal. Between the papillary muscle of the posterior wall and the interventricular septum there was a pocket measuring 1 cm across and 2 cm deep. In the right ventricle the moderator band was enlarged and there was a dysplastic muscle of Luschka. Three defects at different levels were readily evident in the interventricular septum bulged into the right ventricle.

Fig. 1. View from the right hand side to the ventricular septum. Three defects are distinctly to be seen. (SN 959/70 Path. Inst. University Heidelberg)

As seen from the right side of the ventricular septum (Fig. 1):

1. Inferior Defect. This defect, 0.5 cm in diameter, was located between trabecular muscles 2.5 cm from the cardiac apex. Its connection with the pocket of the left ventricle was hidden by the enlarged papillary muscle.

2. Middle Defect. This was in the muscular part of the septum below the crista supraventricularis 3.5 cm from the cardiac apex. It measured 1.5 cm across and was covered by a surgical patch.

3. Superior Defect. This 0.5 cm opening was in the anterior and subinfundibular part of the septum above the crista supraventricularis. Distal to the lower margin of this defect in the anterior septum there was a transverse incision from the surgical cardiotomy.

Summary of Pathological Diagnoses

Three ventricular septal defects at various levels; recent surgical patch-closure of the largest defect in the muscular posterior septum; acute congestive heart failure.

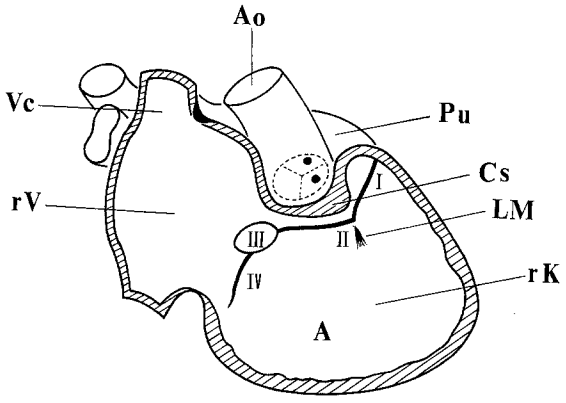

Fig. 2. View from the right of the ventricular septum in the finished heart, illustrating the classification principle of ventricular septum defects. *A* primary ventricular septum. *I* closure zone of the bulbus cushions. *II* raphe of the principle and complementary ridges (bulboauricular ridge). *III* pars membranacea. *IV* fusion zone of the joined endocardial cushions with the principle ridge. *Ao* aorta. *Pu* pulmonalis. *LM* Luschka's muscle. *Cs* crista supraventricularis. *rV* right atrium. *rK* right chamber

Ventricular septal defects are the most common congenital heart defects. According to Becu *et al.* (1956) single defects of the interventricular septum are found in 17% of all congenital cardiac anomalies. Doerr (1967) found the same percentage. Multiple defects of the interventricular septum, however, are comparatively rare. These congenital anomalies are caused by a disturbance in metabolism of the myoepicardial coat (Doerr, 1967). Apparently the type of noxious agent (bacterial, viral, chemical, other causes of metabolic disturbances) is not decisive. What is important is the embryonic heart is injured when septation of the ventricles is taking place. That begins about the 29 th day (at horizon XIV, Streeter, 1945) and ends about the 41 st day (Asami, 1969; Sissman, 1970) or at the XX horizon (Streeter, 1951). The agent causing the injury may act so insidiously the mother may be totally amazed at it. Consequently, pregnancy is usually described as normal, as in the case presented. The recognized principles of classification of interventricular septal defects offer two schemes for analyzing such cases in which several defects are found one above another along the outflow path. The first scheme, derived from that of Rokitansky (1875) and Spitzer (1923) and has been revised by Kl. Goerttler (1960) and improved upon by Doerr (1967). The second scheme was proposed by Becu and Coworkers (1956) and depends on special structures of the right cardiac ventricle.

Since the ventricular septal defects in the heart being presented are located in parts of the septum that embryologically develop at different times it seems only logical to analyze the defects from a purely embryological point of view (Bersch, 1971) (Fig. 2). We know from the literature that the inferior, intertrabecular defect represents a failure of closure of the *primary ventricular septum* (Point A in the classification). The middle defect of the muscular portion of the septum, located below the crista supraventricularis on the right, corresponds to the line where the main septum unites with the opposing bulboauricular septum (Point

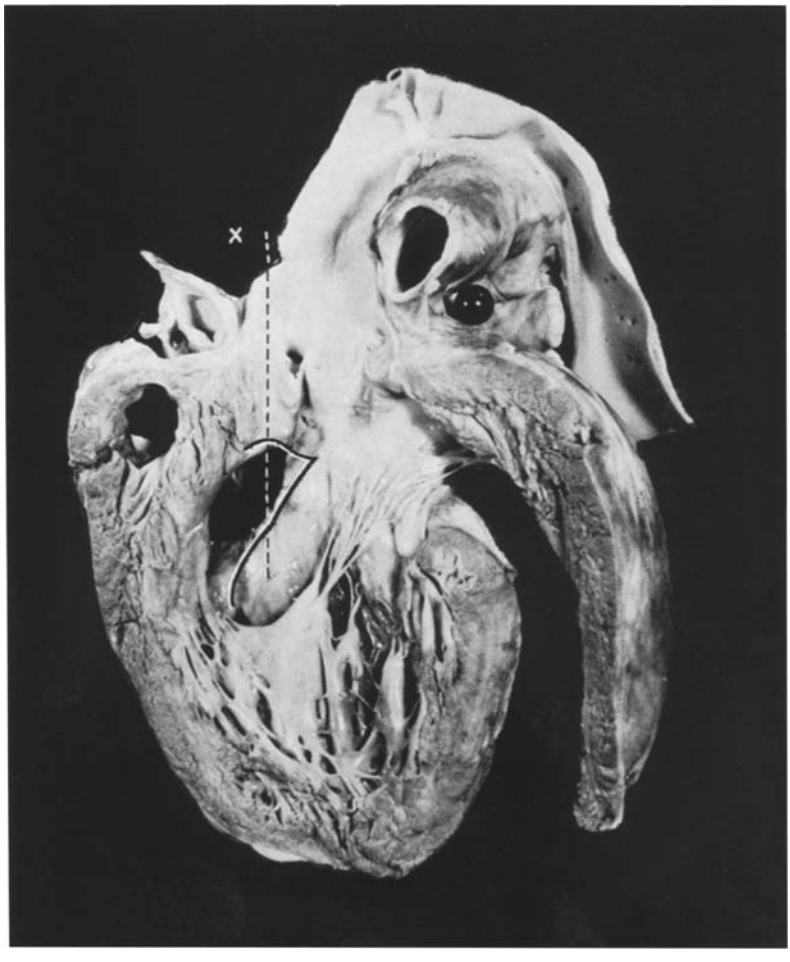

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Fig. 3. a View from the left hand side to the ventricular septum. You only can see the middle and the superior defect. The inferior defect is hidden behind the posterior papillary muscle. The course of the left bundle of the conduction system which bordes the middle defect at top and basis is marked schematically. × line of section of Fig. 3b (SN 959/70 Path. Inst. University Heidelberg). b Microscopical section of the *superior* margin of the middle defect of the ventricular septum. Just below the endocardial limitation of the middle defect you can see the fibers of the conduction system as a small borderline. (Staining: v. G. Enlargement about 100. SN 959/70 Path. Inst. University Heidelberg)

B II in the classification). The third and superior defect of the anterior septum, located subinfundibular and above the crista supraventricularis is in the line of closure of the bulbar septa (Point B I in the classification). If we carefully group all the cardiac septal defects which lie in the secondary zones of closure of the interventricular septum (Point B I–IV of the classification) with the structures that lie along the lines of union we are then able to study how the left bundle of

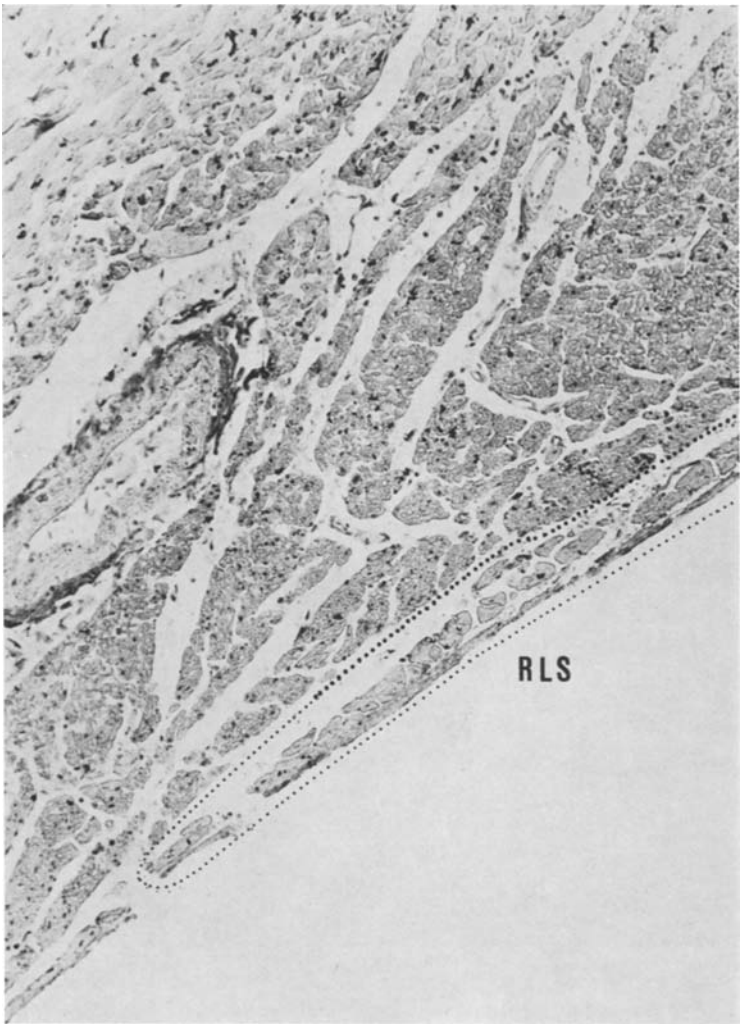

Fig. 3b

the conduction system courses and how it is related to the defects (Bersch, 1972). To do that we prepared histological step-sections of this heart so we could follow the course of the conduction system near the middle and superior defects. The results of our studies are presented in Fig. 3, which schematically portrays the course of the left AV bundle. No fibers of the conduction system were found near the superior defect (the one located beneath the pulmonary artery and above the crista supraventricularis) (Point B I of the classification). These findings support, the assumption that the defect of the closure zone of the bulbar septa is a "pure" defect. The middle defect, in the line along which the main septum unites with the opposing bulboauricular septum (Point B II of the classification) represents the most common place for a ventricular septal defect to develop (Becu *et al.*, 1956;

Kl. Goerttler, 1960; Doerr, 1967, 1970). In contrast to the superior defect this defect is bounded along its upper and lower margins by fibers of the conduction system. Consequently the opening is *not* a defect in the region of the bulbus as the topic would lead us to expect. Rather, the fibers coursing above the defect (Fig. 3b) show:

1. The zone of union in unchanged.

2. The septal defect lies near the main septum or the primary interventricular septum, since the fibers, as ventral extensions of the left branch of the AV connecting bundle, run along the line of union of the main septum with the opposing bulboauricular septum (Bersch, 1972).

By studing how the fibers of the conduction system course, we have attempted to confirm the embryological principle of classification of cardiac anomalies with multiple ventricular septal defects.

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