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Editorial

Is an interatrial communication the same as an atrial septal defect?

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It is, perhaps, unfortunate that those using Anglo-saxon tongues, as opposed to romantic languages such as French, Spanish, Italian, or Portuguese, should describe holes permitting shunting between the atrial chambers as septal defects rather than interatrial communications. This is because, of necessity, the understanding of deficiencies of the atrial septum is entirely dependent on the definition of the structures considered to represent the atrial septum.[1] Interatrial communications, on the other hand, describe all those holes that provide the facility for shunting between the atrial chambers. Those with limited interest in congenital cardiac malformations may well wonder why such details are of functional significance. The reason is that, of the five phenotypic arrangement known to produce interatrial communication, only 2 are due to deficiencies of the atrial septum. The most common defect is due to deficiency of the flap valve of the oval foramen, either because the flap valve is perforate, or else because the valve is of insufficient size to overlap the rims of the oval foramen. These defects are known as "secundum" defects, even though they are due to abnormal formation of the primary atrial septum, which forms the flap valve of the oval foramen.[1] A second true septal defect, albeit very rare, involves the antero-inferior muscular buttress from which the flap valve is hinged. This is the so-called vestibular defect.[2] The remaining holes that produce the potential for interatrial shunting are all outside the confines of the oval fossa, and hence are interatrial communications rather than septal defects. Of these three, the most common is the "ostium primum" defect, now well recognised as being an atrioventricular rather than an atrial septal defect. Its phenotypic feature is the commonality of the atrioventricular junction, with the left valve being trifoliate, rather than representing a "cleft" mitral valve.[3] The rarest interatrial communication is the coronary sinus defect, produced by absence of the walls that, in the normal heart, separate the cavities of the coronary sinus and left atrium.[4] It is the third defect that is perhaps the most interesting. This is the sinus venosus defect, positioned most frequently in the mouth of the superior caval vein,[5] but less frequently in relation to the inferior caval vein.[6] In this issue of the journal, the group from Sao Paulo describe the finding of such an inferior sinus venosus defect co-existing, in an adult patient, with a large defect within the oval fossa.[7] As they explain, the feature of the sinus venosus defect is the anomalous connections of the right inferior pulmonary veins, which connect anomalously to the inferior caval vein whilst retaining their connection to the left atrium.

The morphogenesis of sinus venosus defects has been controversial. For some time, we believed ourselves that the essence of such defects was overriding of the rims of the oval fossa by one or other of the caval veins. This explanation was shown to be invalid, however, when we encountered unequivocal cases of the superior defect in which the superior caval vein was connected exclusively to the right atrium.[8] As we showed in this most recent publication, in reality the lesions are veno-venous malformations, rather than septal defects.[8] We also showed that the alternative explanation, that of "unroofing" of the right pulmonary veins,[9] was invalid simply because, in the normal heart, there is no party wall between the right pulmonary veins and the right atrium. The case described here provides additional evidence that the true essence of the sinus

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venosus defect is anomalous connection of one or more pulmonary veins to a systemic venous channel, with the anomalous pulmonary venous structures retaining their left atrial connection, thus producing an extracardiac conduit which provides the potential for interatrial shunting.

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