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## **Defining the morphologic phenotypes of atrial septal defects and interatrial communications**

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### **Abstract**

True atrial septal defects are limited to deficiencies totally within the confines of the oval fossa and its antero-inferior rim. Other communications between the two atriums, such as the superior or inferior sinus venosus defects, coronary sinus defect, and the ostium primum defect, are less frequently seen. They represent interatrial communications rather than septal defects, since the communication between the atrial chambers is outside the confines of the true atrial septum. The ostium primum defect, for example, has all the characteristics of an atrioventricular septal defect, existing only in the setting of a common atrioventricular junction. The unifying physiological feature of all these variants, whether their morphology is that of a defect within the area of the oval fossa, or an opening elsewhere within the atrium, is that mixing of the systemic and pulmonary blood occurs at atrial level. In this review, we emphasise the distinction between true atrial septal defects and defects which result in communications between the atriums, underlining the phenotypic characteristics, along with the notable morphological features that are of significance in current strategies for clinical management.

**MeSH:** heart defects, congenital, atrial septal defect, interatrial communication, morphology

### **Article**

#### **Assessing defects between the atriums**

Communications between the atrial chambers represent one of the most common heart defects, accounting for about one-tenth of all congenital

cardiac lesions.<sup>1,2</sup> In adults who have congenital cardiac disease, approximately one-third will have an atrial septal defect, and this occurs in females three times as often as in males.<sup>3</sup> Nowadays, various techniques are used for closure of holes between the atriums. These include minimal access surgery, and percutaneous closure using a device inserted on a catheter. These advances in treatment have increased the need for accurate assessment of the defect, not just of its size, but also its morphology and spatial relationship within the atrium, before commencing treatment.<sup>4-6</sup> Several techniques are used to image and size atrial septal defects. These include cross sectional echocardiography, intracardiac ultrasound, magnetic resonance imaging, and balloon sizing during cardiac catheterisation. Currently, the most commonly used imaging technique is cross-sectional echocardiography. The use of this technique in centers today permits the detection of more than 90% of all interatrial communications,<sup>7</sup> using transthoracic and transoesophageal echocardiography to view the defect in various planes. Although extremely efficient, this method may not fully reflect the true size, or provide a full picture of the entire margins of the defect throughout the cardiac cycle. This is essential information, as the maximal diameter of the defect, its precise dimensions and location, are crucial indicators to select patients either for interventional or surgical closure. Proper assessment is crucial to determine the strategy for treatment, as some defects are unpredictable, and can present as small insignificant defects, but over time they can develop into haemodynamically important conditions.<sup>8</sup> In one study conducted over an eight-year period, two thirds of atrial septal defects within the oval fossa were found to enlarge over time, so becoming inappropriate for transcatheter closure.<sup>9</sup> There have been further technological advances in imaging with the introduction of 3-dimensional echocardiography. This imaging technique allows the entire morphology of the defect to be observed in real time, and therefore permits study of the pathophysiology of atrial shunting.<sup>10,11</sup> This technique provides a more accurate description of the varying dynamic morphology, dimensions, and spatial relationship of the interatrial communications.<sup>12,13</sup> This is important, as a recent clinical study, which measured the size of the defect, found that this did not always correlate to the degree of shunting, or the resulting clinical symptoms.<sup>14</sup> Further advances using magnetic resonance technology are now being trailed in animal studies as an alternative to using ionizing radiation when closing a patent oval fossa.<sup>15</sup> But in order to appreciate the spectrum of phenotypes responsible for interatrial shunting, in relation to their clinical management, it is first essential to appreciate the normal septal anatomy.

### **Development of the normal atrial septum**

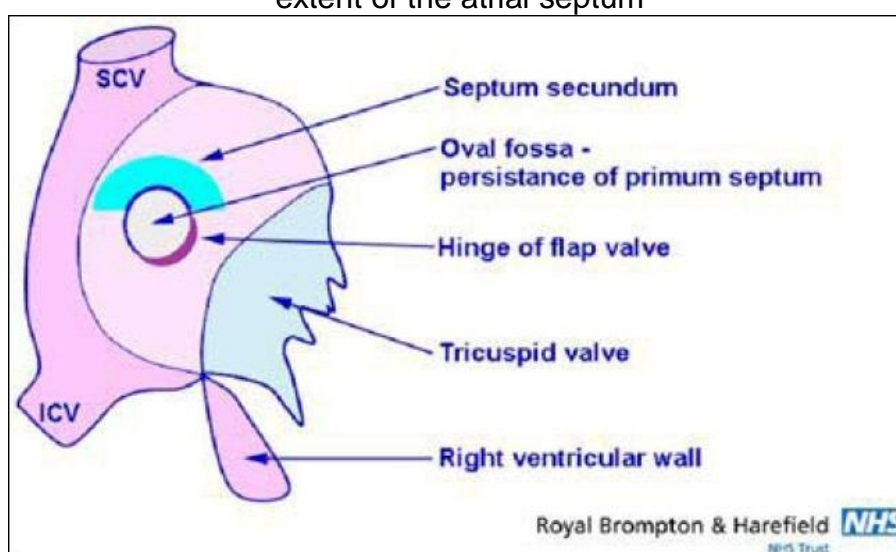
The development of the normal atrial septum occurs following the initial looping of the heart. In the embryonic heart, the normal atrial septum, and the surrounding atrial structures, are formed from several embryological tissue components that develop, remodel and fuse in the correct sequence.

As the initial step in septation, a ridge of tissue develops from the superior aspect of the primary atrial component of the heart tube. This ridge is the primary septum (septum primum), and its the leading edge is covered by

cushion-like mesenchymal tissue that is continuous over the dorsal mesocardium. This dividing crest of tissue is part of the atrial chamber expressing genes demonstrating morphologically leftness.<sup>16</sup> As it grows into the atrial cavity, it extends down towards the endocardial cushions that are developing concomitantly within the atrioventricular canal. Normal septal development also involves incorporation of another mass of tissue derived from the dorsal mesocardium. This is known as the vestibular spine (spina vestibuli), and it, too, carries on its leading edge a mesenchymal cap. As the primary septum approaches the atrioventricular endocardial cushions, the various mesenchymal structures fuse together.<sup>17</sup> The mass derived from the vestibular spine then muscularises, eventually forming the prominent infero-anterior border of the oval foramen.<sup>18</sup> During the process of development the ventricular septum also ‘moves’ up towards the endocardial cushions, resulting in the septation of the ventricular chambers.<sup>19</sup>

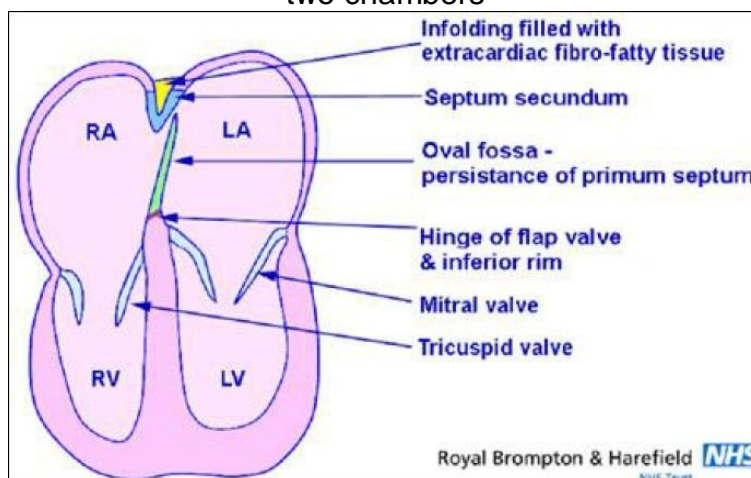
Subsequent to the fusion between the primary septum and the endocardial cushions of the atrioventricular canal, the upper part of the primary septum disintegrates to form the ‘ostium secundum’. The remaining part of the primary septum becomes the flap valve of the oval fossa. This flap valve, along with the muscularised antero-inferior rim, forms the true septum that separates the cavities of the atrial chambers. Only after integration of the pulmonary veins into the left atrium do the superior walls of the two atriums ‘in角度’, creating the so-called “septum secundum” in the superior portion of the atriums. The flap valve overlaps, but is not completely adherent to, the rims of this superior atrial fold, also known as Waterston's or Sonderggard's groove, providing a passage during fetal life for blood to pass from the right to the left atrium (Figure 1). In postnatal life, this deep superior interatrial fold becomes filled with extracardiac fibro-fatty tissue (Figure 2).

Figure 1 Diagrammatic representation viewing the oval fossa from the right atrial aspect. The oval fossa flap valve and the immediate rim is the true extent of the atrial septum



SCV, superior caval vein. ICV, inferior caval vein.

Figure 2 Diagrammatic representation of the heart in 4-chamber section. The flap valve overlaps the superiorly infolded walls of the atriums, partitioning the two chambers

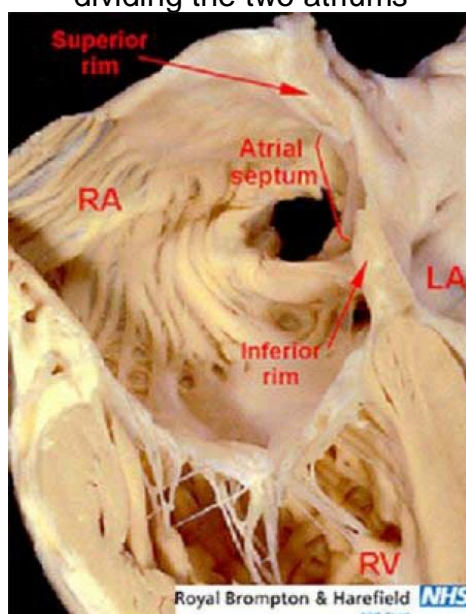


RA, right atrium. RV, right ventricle. LA, left atrium. LV, left ventricle

### The normal postnatal heart

In the definitive postnatal heart, when the wall separating the atriums is viewed from its right atrial aspect, the septal aspect, at first sight, seems extensive. The atrial septum, nonetheless, is best defined as the tissues which directly separate the atrial cavities, and which can be removed without exiting the heart. When viewed in the light of this definition, the septum is confined to the thin flap of fibromuscular valvar tissue which forms the floor of the oval fossa, along with the immediate infero-anterior muscular rims of the fossa derived from the vestibular spine (Figure 3).

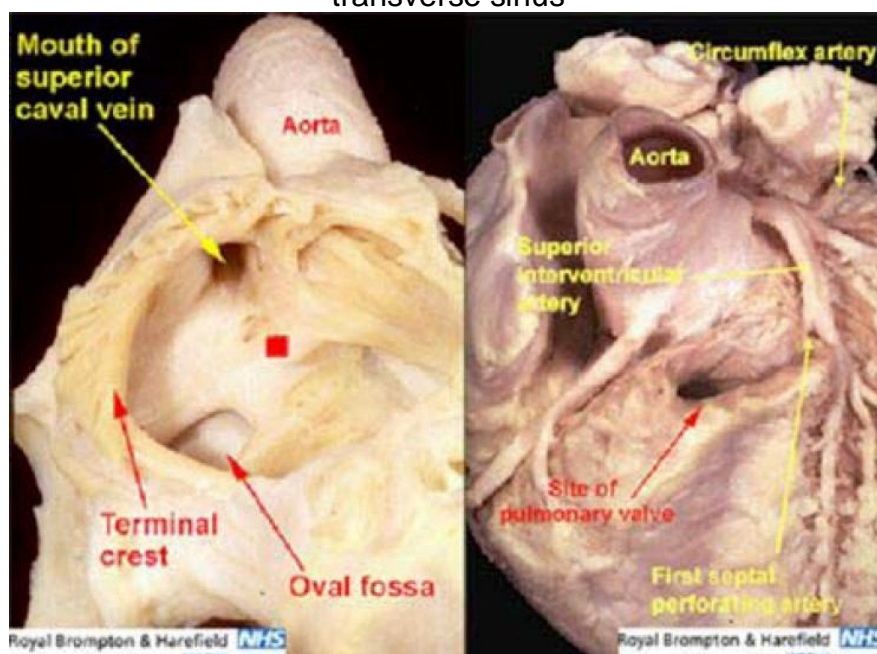
Figure 3 Close-up four chamber view of the RA and RV, showing the muscular superior and inferior rims, and the flap valve of the oval fossa dividing the two atriums



RA, right atrium. RV, right ventricle.  
LA, left atrium

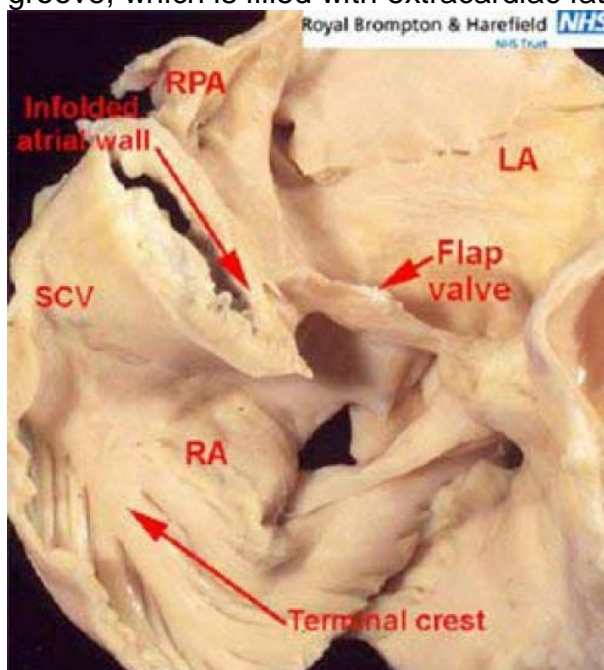
Thus, the septal area is only a small part of the wall dividing the atriums. The aortic mound, in contrast, which is to the right of the oval fossa when observed from the right atrial aspect, and which seems to represent an apparently solid muscular structure, is part of the external wall of the heart. Passage of an instrument through this area does not take one into the left atrium, but rather into the transverse sinus of the pericardial cavity, in front of the bulging right coronary sinus at the base of the aortic root (Figure 4).

Figure 4 Figure 4A: Anterior view of the right atrium, showing the relationship of the aortic mound (red square) to the oval fossa. Figure 4B: A needle passing through the aortic mound (red square) would exit the heart into the transverse sinus



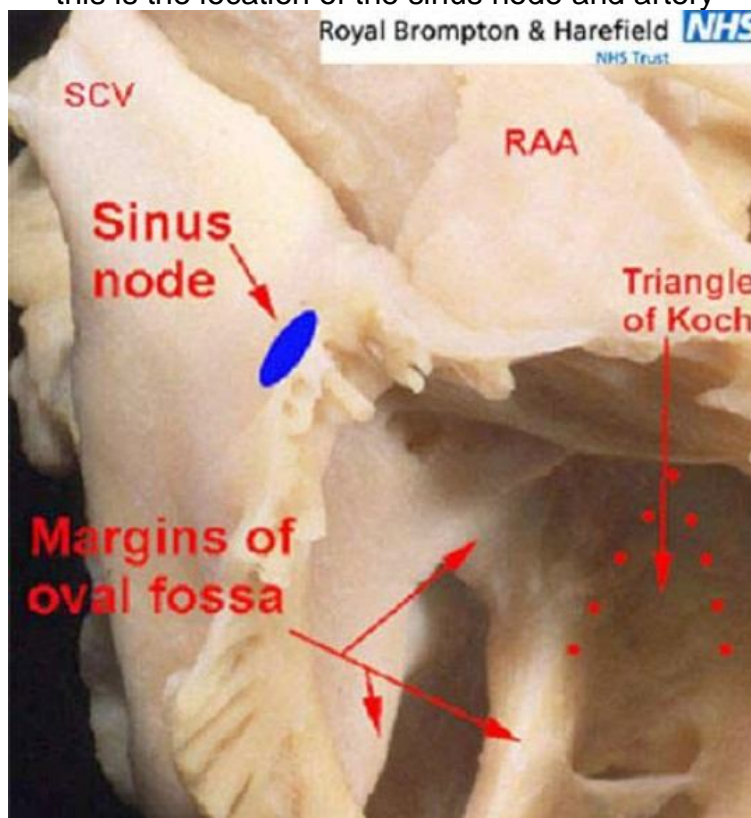
Similarly, passing a needle from the right to left atrial chambers superiorly to the oval fossa also passes through extracardiac tissue, as this part of the dividing wall is in reality an infolding of the right atrial wall and hence is not septal (Figure 5). In adult life, this fold is filled with fatty extracardiac tissue. It is within this area that the sinus nodal artery usually takes its course (Figure 6).

Figure 5 Four chamber view showing the infolded right atrial wall, Waterstons groove, which is filled with extracardiac fat.



RPA, right pulmonary artery. SCV, superior caval vein.  
RA, right atrium. LA, left atrium

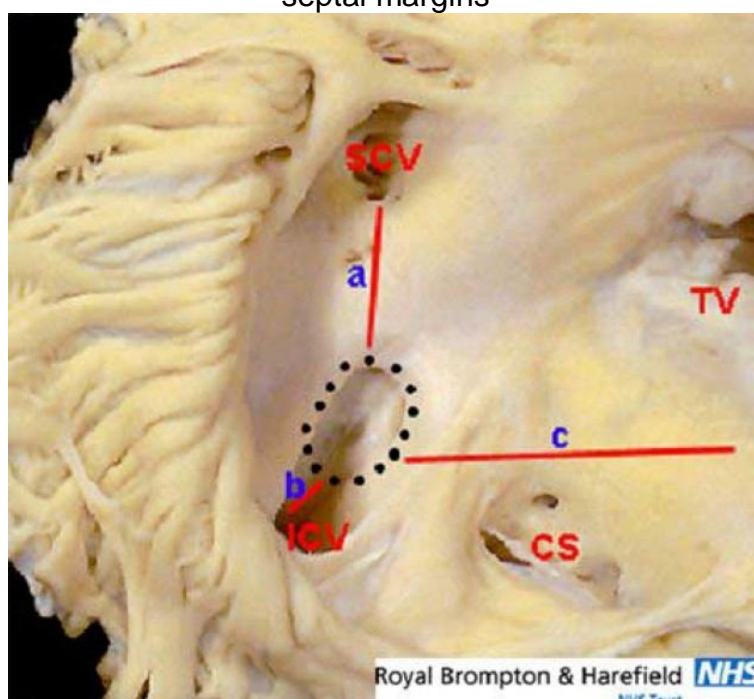
Figure 6 Anterior view showing the junction between the RAA and the SCV, this is the location of the sinus node and artery



SCV, superior caval vein. RAA, right atrial appendage

Morphologically, the distinct rims of the oval fossa are related to other important structures within the atrium. These can be viewed echocardiographically. The superior margin extends from the superior edge of the atrial septal defect towards the attachment of the superior caval vein within the right atrium. As emphasised, this superior rim is essentially an infolding of the muscular atrial walls. Progressing towards the postero-inferior rim of the defect, we find the attachment of the inferior caval vein within the right atrium. The remaining border is the important anterior margin. This separates the margin of the oval fossa from the annulus of the tricuspid valve and the orifice of the coronary sinus (Figure 7). Full interrogation of the nature of these muscular borders of the oval fossa is crucial when assessing whether an interventional device can safely be fitted to close off a defect.

Figure 7 Right atrial view showing the oval fossa (black dots) surrounded by septal margins



a: superior rim. b: postero-inferior rim. c: anterior rim. ICV, inferior caval vein. SCV, superior caval vein. CS, coronary sinus. TV, tricuspid valve

## Defects within the Confines of the Atrial Septum

### Patent oval fossa, and secundum defects within the fossa

In one-quarter to one-third of the normal population, the atrial septum does not close completely in the neonatal period. In these cases, the upper margin of the flap valve overlaps the infolded antero-superior rim of the oval fossa, but does not become fused to it. Because of the failure of fusion, should right atrial pressure be higher than left, there is the potential for communication between the atriums within the region of the oval fossa. It is not due to any deficiency of the intrinsic septal structures, but it is created by a failure of the flap valve fully to adhere to the entirety of the rim. This lack of adhesion will permit a probe to be passed obliquely from the right to the left atrium. Usually,

probe patency is not physiologically significant, and is often an incidental finding at post-mortem. The patent flap valve will permit inter-atrial shunting only occur when right atrial systemic pressure is higher than that in the left atrium.

True defects of the atrial septum, of necessity confined within the bounds of the oval fossa, are often referred to as 'secundum' defects. The defect, nonetheless, is a deficiency of the floor of the oval fossa, and this floor is derived from the primary atrial septum. Thus, the defects are 'ostium secundum' defects, and not deficiencies of the secondary atrial septum. As we have shown, the secondary "septum" is no more than an infolding of the atrial roof. Such defects within the confines of the fossa account for over three-quarters of all holes between the atriums. In a study of 160,480 live births, atrial septal defects were found to account for one-twentieth of all cases of congenital heart disease diagnosed in infancy.<sup>20</sup> The occurrence of congenital heart disease in a total of 43 studies was reviewed recently, and the overall incidence of atrial septal defects was found to be 3.4/10,000 of live births.<sup>21</sup> In adults with a congenital heart defect, 'secundum' atrial septal defects comprise nearly a third of all cases.<sup>3</sup>

The probability of spontaneous closure of an isolated atrial septal defect in patients below the age of 5 years is high, with closure commonly occurring in up to four-fifths of the small to moderate defects that are diagnosed within the first three months of life.<sup>22</sup> Spontaneous obstruction of the hole has also been observed beyond infancy. In one study, spontaneous closure was found in three-fifths of the population studied after the age of 18 months, and in total two-fifths of all the patients studied exhibited a natural closure after the age of five years.<sup>23</sup> The mechanism of spontaneous closure is unknown. Small atrial septal defects can remain completely asymptomatic and hemodynamically insignificant throughout life.

Morphologically, there are notable variations in the structure of these atrial septal defects. The differences impact on transcatheter closure. Intervention may not be possible if the defect is too large, or if there is a lack of suitable rims around the entire circumference to provide anchorage for a device. Other contraindications included the defect being displaced towards the posterior wall, and its proximity to the entrance of the superior or inferior caval veins. Also, if the eustachian valve is thick, this structure can obscure the postero-inferior rim of the oval fossa.<sup>24</sup> When examined morphologically, seven-tenths of defects were centrally located defect within the confines of the oval fossa.<sup>25</sup> Even within a group having these characteristics, the borders of the defect in two-fifths of the cases were deemed insufficient to provide firm anchorage for an occluding device.<sup>26</sup>

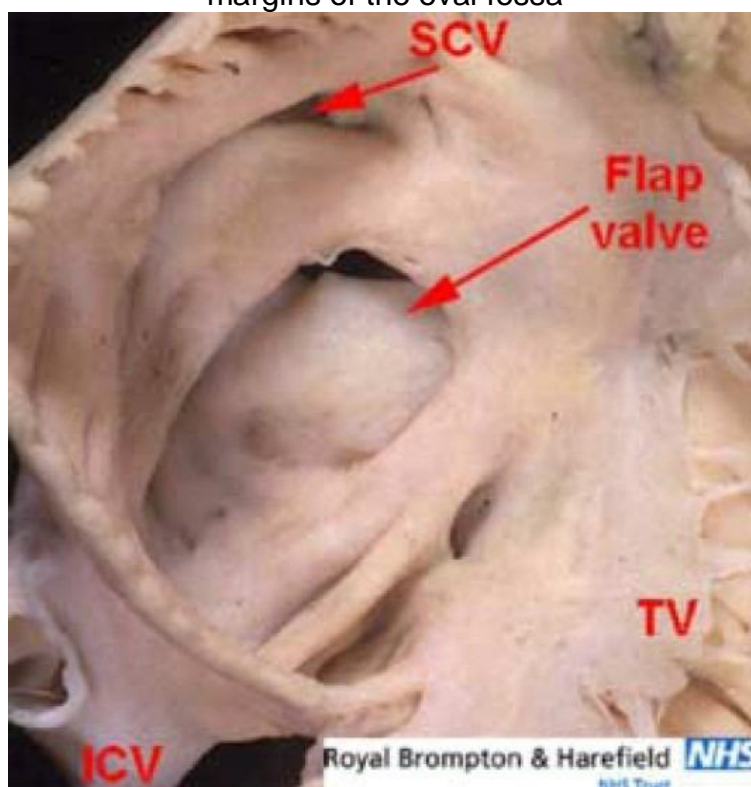
The extent of deficiency of the flap valve can produce a spectrum of morphology, ranging from the flap valve failing to cover fully the oval fossa completely (Figure 8), to its complete absence (Figure 9). Absence of the flap valve altogether, along with effacement of the rim, leads to a physiologically functional 'common atrium'. The morphology of the floor itself can also be variable, with some flap valves being very thin and membranous, composed of connective tissues, to others that have integration of myocardium and so become thick and muscular. The Rashkind procedure for



atrial septostomy will clearly be more difficult if a thicker and more muscular floor of the oval fossa is encountered.

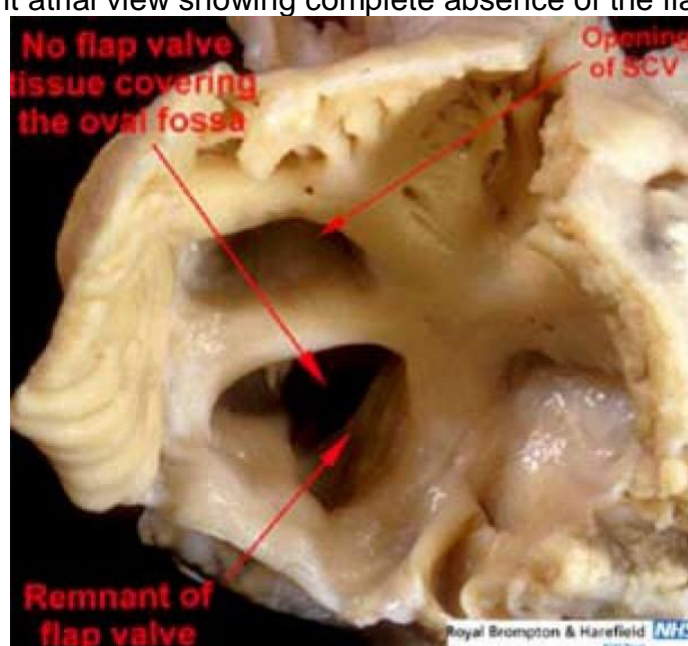
Once the defect is assessed, and it is determined that closure by device is recommended, then in ideal cases, this is a relatively straightforward procedure (Figure 10).

Figure 8 Right atrial view showing flap valve tissue that fails to fully cover the margins of the oval fossa



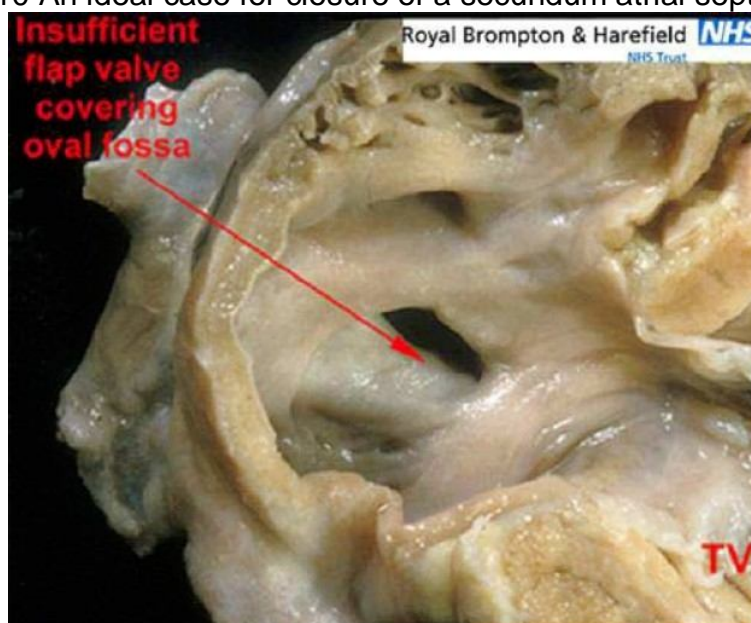
ICV, inferior caval vein. SCV, superior caval vein. TV, tricuspid valve

Figure 9 Right atrial view showing complete absence of the flap valve tissue



SCV, superior caval vein

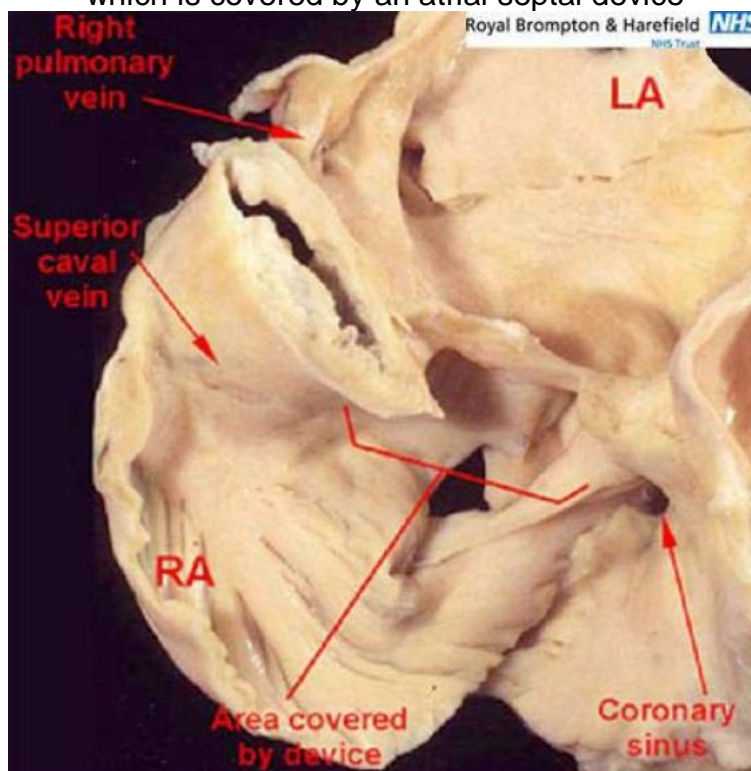
Figure 10 An ideal case for closure of a secundum atrial septal defect



TV, tricuspid valve

Contraindications to closure include a large defect, even when a suitably large device is available. If the defect is unduly large, the device is likely to be adjacent to important structures with the atriums (Figure 11).

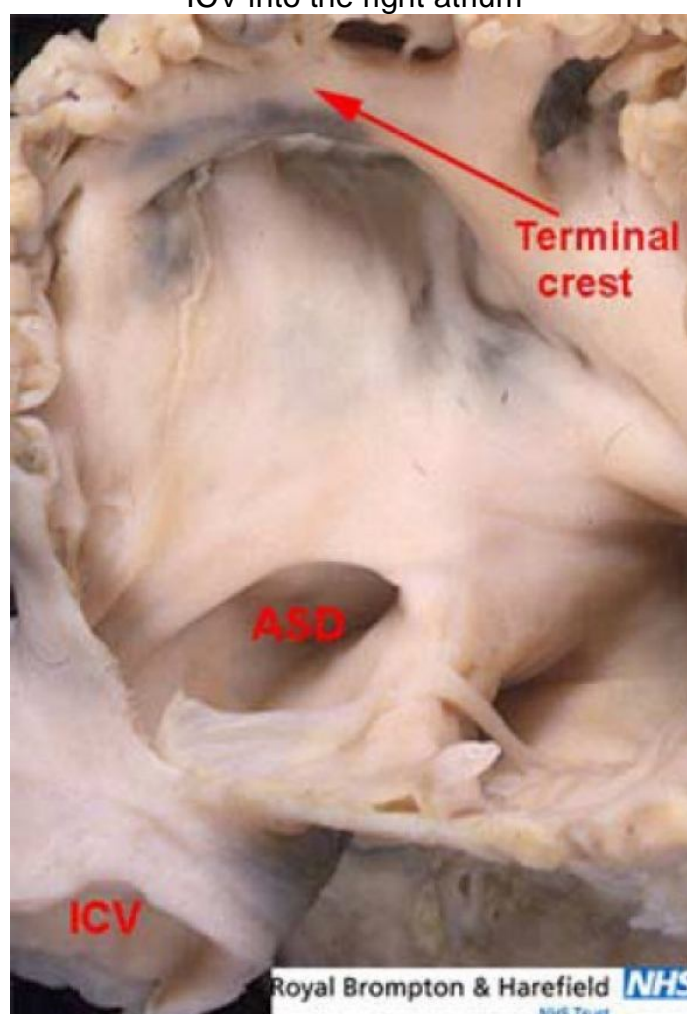
Figure 11 Shows the proximity of the superior caval vein, the coronary sinus in the right atrium and the right pulmonary vein in the left atrium to the area which is covered by an atrial septal device



LA, left atrium. RA, right atrium

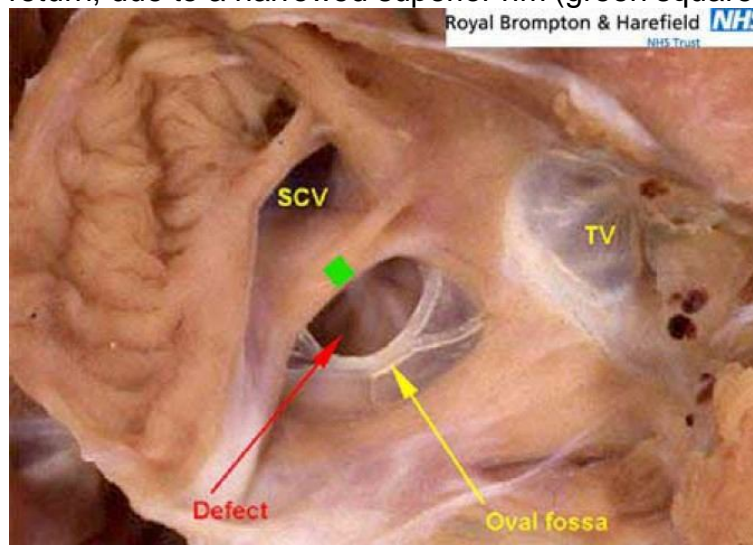
The final position of the device is also of crucial importance, irrespective of the size of the defect, as its edges could impinge inferiorly in the right atrium on the coronary sinus, the atrioventricular nodal area, and the inferior caval vein (Figure 12), or the device could encroach on the leaflets of the mitral valve in the left atrium. In one study, in one-third of cases the narrowest boarder of the defect was found to be between the oval fossa and the aortic mound, at the antero-superior rim.<sup>27</sup> The same study showed the superior rim to be furthest from the defect. A short rim in this position could result in the right upper and lower pulmonary veins being occluded on the left atrial aspect, or the superior caval vein being obscured on the right atrial aspect (Figure 13). Holes can occur at various locations within the flap valve itself (Figures 14,15). In a recent review of post-mortem specimens, over half were found to have a fenestrated flap valve.<sup>27</sup> The degree of fenestration can vary greatly. If widespread, the floor of the fossa can have a fishnet-like appearance, with the tissue becoming quite extensive.

Figure 12 This inferiorly located atrial septal defect is near the entrance of the ICV into the right atrium



ICV, inferior caval vein. ASD, atrial septal defect

Figure 13 A device placed to close this defect may occlude the SCV venous return, due to a narrowed superior rim (green square)



TV, tricuspid valve. SCV, superior caval vein

Figure 14 View of the right atrium showing fenestrations within the flap valve of the oval fossa

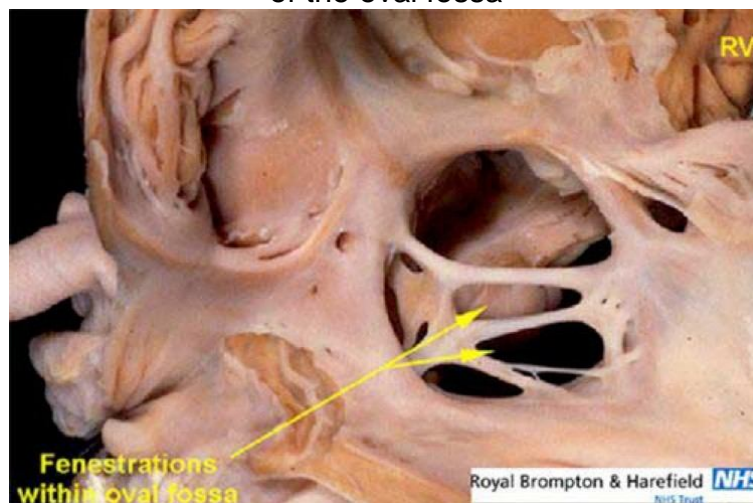
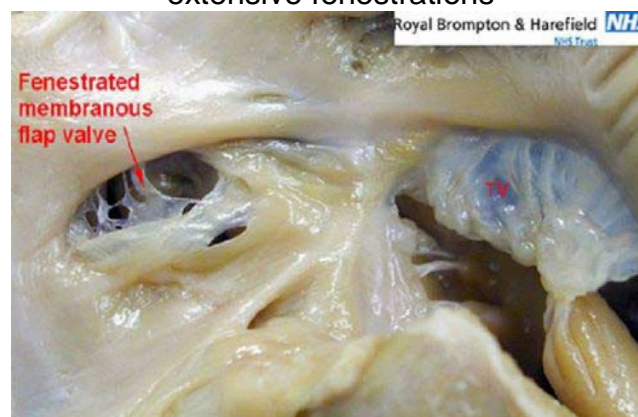
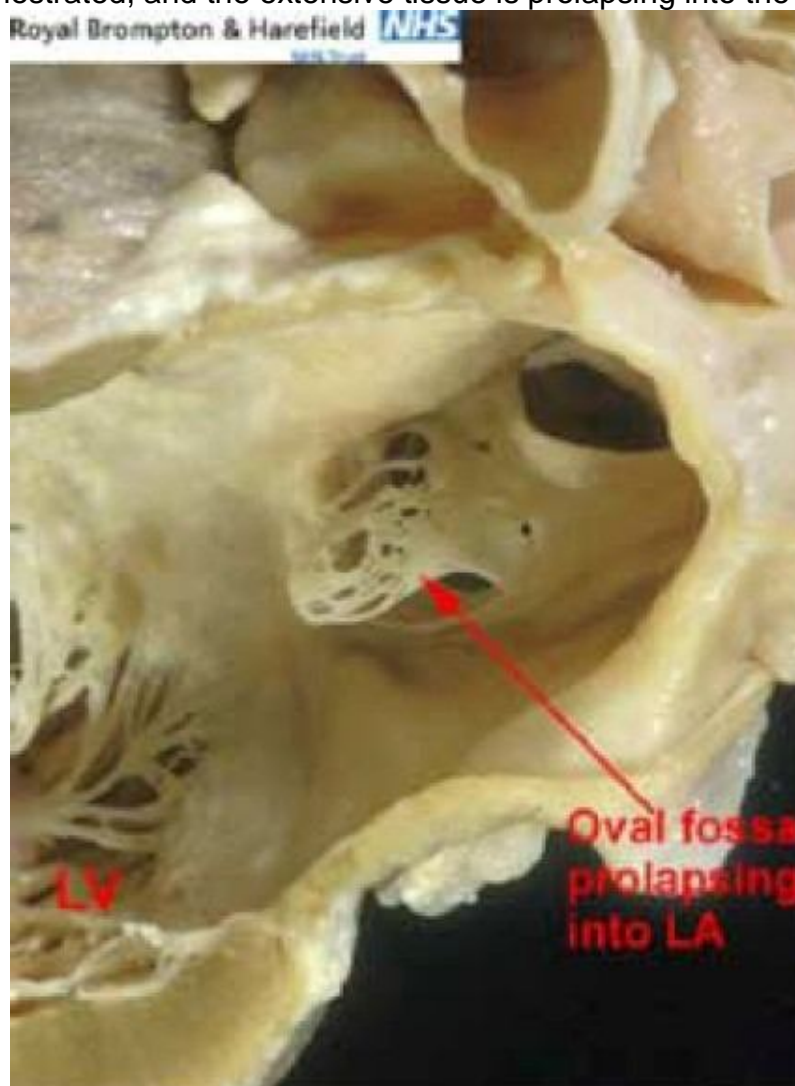


Figure 15 Right atrial view in another heart, the oval fossa is fine and shows extensive fenestrations



TV, tricuspid valve. CS, coronary sinus

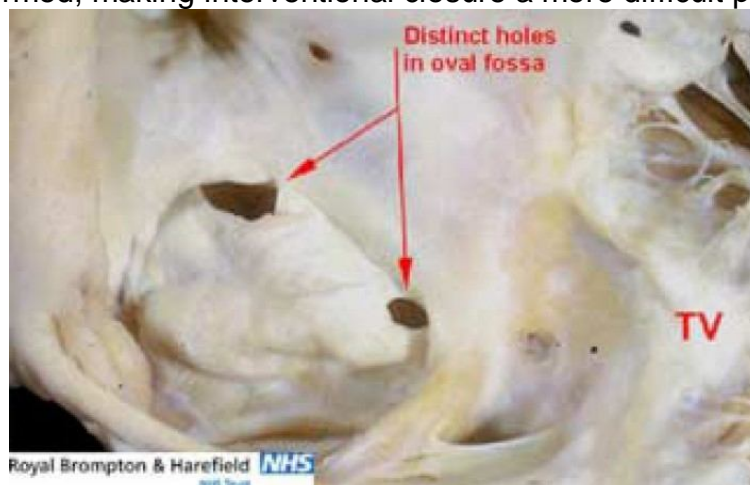
Figure 16 Seen from the left atrial aspect the flap valve of the oval fossa is highly fenestrated, and the extensive tissue is prolapsing into the left atrium



LV, left ventricle. LA, left atrium

In these circumstances, the flap valve can take the appearance of a windsock blowing into the left atrium (Figure 16). Such extensive flap valves also have the potential to become aneurysmal in the opposite direction, producing a left-to-right shunt with right ventricular overload.<sup>25</sup> Conversely, a few widely separated holes may complicate the strategies for treatment (Figure 17).

Figure 17 A spectrum of deficiencies can affect the oval fossa flap valve, giving rise to various degrees of fenestrations. In this case two separate holes have formed, making interventional closure a more difficult procedure



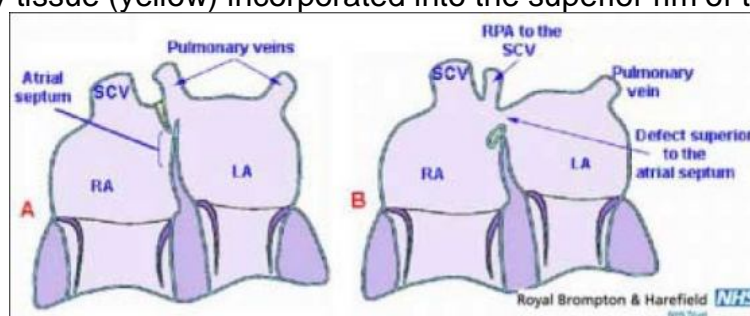
TV, tricuspid valve

## Defects outside the Confines of the Atrial Septum

### Superior and Inferior sinus venosus defects

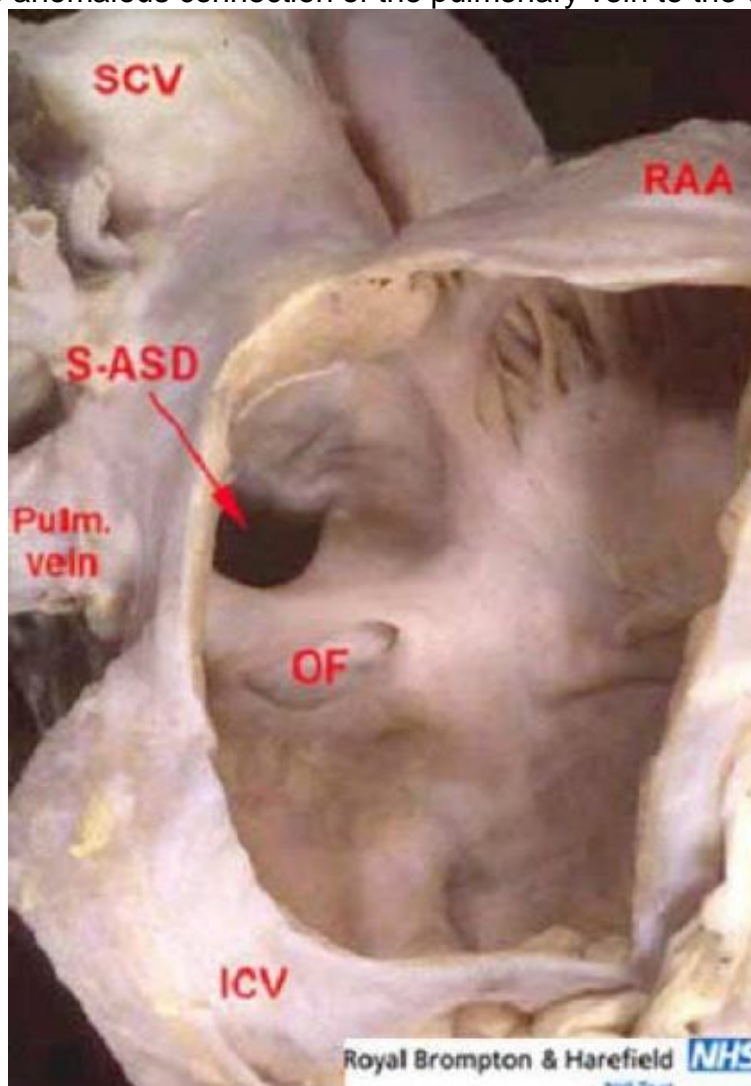
These defects are both examples of interatrial communications in which the atriums communicate through a channel outside the boundaries of the true atrial septum (Figure 18). In cases of superior sinus venosus defect, the hole is located superiorly to the oval fossa, which can itself be intact or deficient. In the typical morphology seen in hearts with a superior sinus venosus defect, the superior caval vein usually overrides the crest of the defect. The caval channel then has biatrial connections, opening into the right and left atriums.<sup>28</sup> The phenotypic feature of the lesion is the presence of the defect outside the confines of the normally formed oval fossa (Figure 19). Indeed, it is possible to pass a probe through the extracardiac tissue found within the intact superior muscular rim of the oval fossa, which forms the inferior rim of the defect itself.

Figure 18 A) The atrial septum in the normal heart. The interatrial infolding is filled with extracardiac fibro-fatty tissue (yellow). B) In hearts with a superior sinus venosus defect the defect is superior to the oval fossa. Extracardiac fibro-fatty tissue (yellow) incorporated into the superior rim of the septum



SCV, superior caval vein. RPA, right pulmonary artery. RA, right atrium. LA, left atrium

Figure 19 The defect overrides the superior rim of the oval fossa, resulting in the SCV having direct communication to both the right and left atriums. Note the anomalous connection of the pulmonary vein to the SCV



S-ASD, superior sinus venosus defect.  
OF, oval fossa. RAA, right atrial  
appendage. Pulm. vein, pulmonary  
vein. ICV, inferior caval vein. SCV,  
superior caval vein

The cases with abnormal pulmonary venous drainage are unsuitable for device closure, and can be complicated to close at surgery. The surgeon must exercise caution and avoid the junction between the superior caval vein and the right atrial appendage, as this is the location of the sinus node (Figure 20). Additionally, the area around the junction of the superior caval vein and the terminal groove must be avoided, as this is the location of the sinus nodal artery.

Inferior sinus venosus defects are uncommon, and occur at the mouth of the inferior caval vein, near the opening of the coronary sinus, but the oval fossa retains its discrete muscular border. These are often difficult to diagnose echocardiographically, since they can be mistaken for large defects of the

oval fossa which extend back into the atrium so that the inferior caval vein overrides the entrance of the oval fossa, thus producing a biatrial communication in the setting of an oval fossa defect.<sup>29</sup> As with all these cases with interatrial shunting, the phenotypic feature of the inferior sinus venosus defect is that communication takes place outside the confines of the oval fossa.

Figure 20 Anterior view of the right atrium, showing the location of the sinus node and artery



RAA, right atrial appendage. SCV, superior caval vein

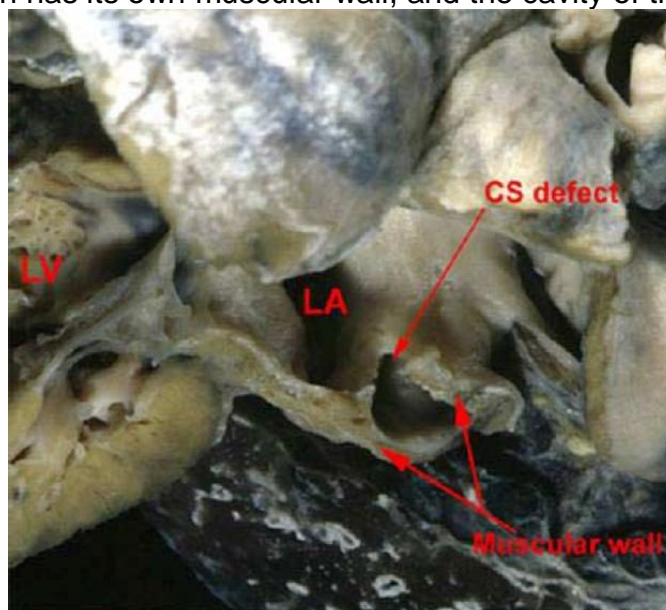
### Coronary sinus defects

The coronary sinus is a venous channel that is located within the left atrioventricular groove, above the annulus of the mitral valve. Distally located tributaries join to convey the deoxygenated blood back to the right atrium. Defects within the wall of the coronary sinus are a rarity. They are usually found associated with anomalous connection of the left superior caval vein to the roof of the left atrium, which enters between the right pulmonary veins and the left atrial appendage. In the normal heart, the muscular wall of the coronary sinus is separated by extracardiac tissue from the inferior wall of the left atrium. Thus, the coronary sinus is an independent venous structure with



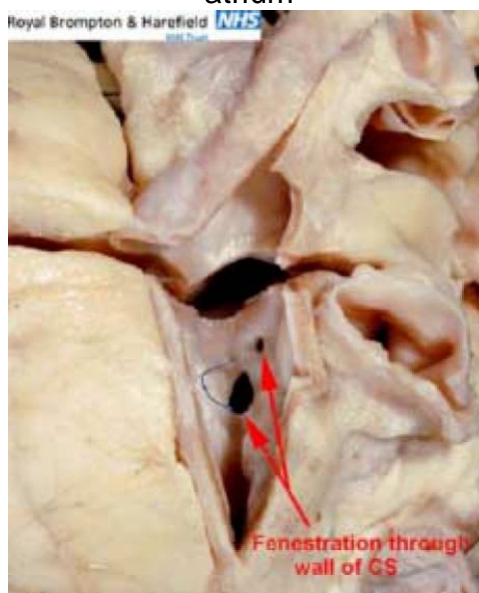
a distinct wall from the atrium. Developmentally, therefore, the existence of this type of interatrial defect is remarkable. In essence, the defect necessitates breakdown not only of the wall of the discrete venous channel, but also partial dissolution of the wall of the adjacent left atrium. The degree of disintegration of the two walls can vary widely, from small distinct fenestrations (Figures 21,22), to complete ‘unroofing’ of both walls, producing complete mixing of the venous deoxygenated blood and the oxygenated blood within the left atrium (Figure 23). The key phenotypic feature is again the presence of the defect outside the confines of the oval fossa.

Figure 21 View of the left atrium showing a fenestration between the coronary sinus, which has its own muscular wall, and the cavity of the left atrium



LV, left ventricle. LA, left atrium. CS, coronary sinus

Figure 22 Posterior view of the heart showing a coronary sinus defect, where parts of the wall are absent allowing discrete communication into the left atrium

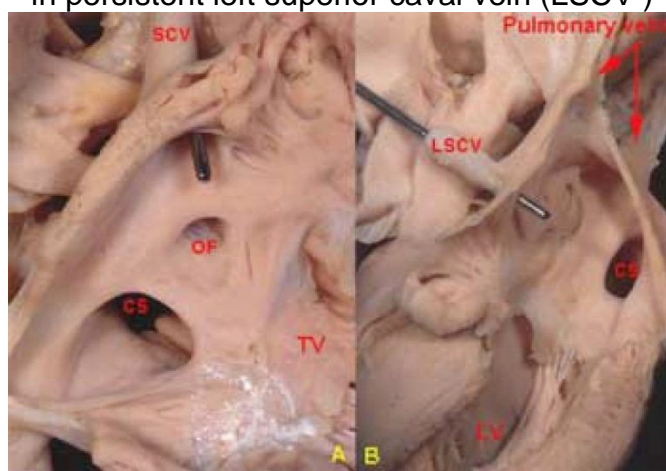


LA, left atrium. CS, coronary sinus

### The Ostium Primum Defect

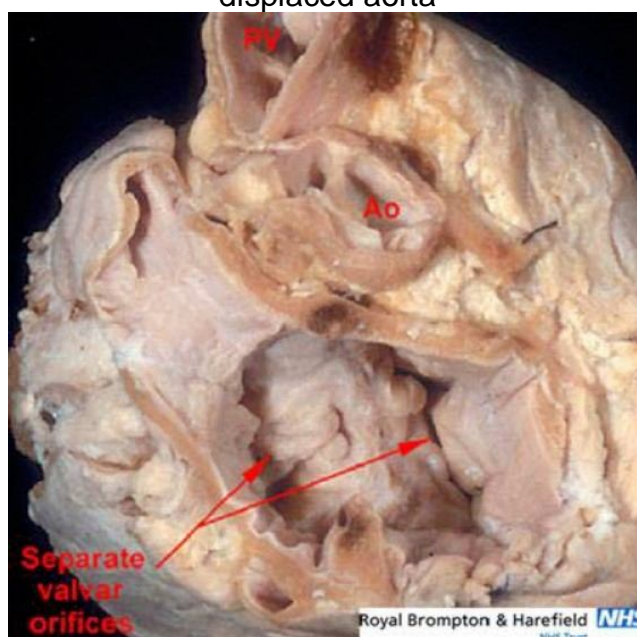
Another lesion producing an interatrial communication is the “ostium primum” variant of atrioventricular septal defect. But here the phenotypic feature in these hearts is the presence of a common atrioventricular junction, albeit with separate valvar orifices into the right and left ventricles (Figures 24).

Figure 23 A) Right atrial view displays an enlarged orifice to the coronary sinus. (Probe in SCV) B) Left atrial view of same heart, there is no coronary sinus channel running within the posterior wall of the left atrium. Instead the coronary sinus opens directly from the right atrium into the left atrium. Probe in persistent left superior caval vein (LSCV)



LV, left ventricle. CS, coronary sinus. OF, oval fossa. TV, tricuspid valve. SCV, superior caval vein

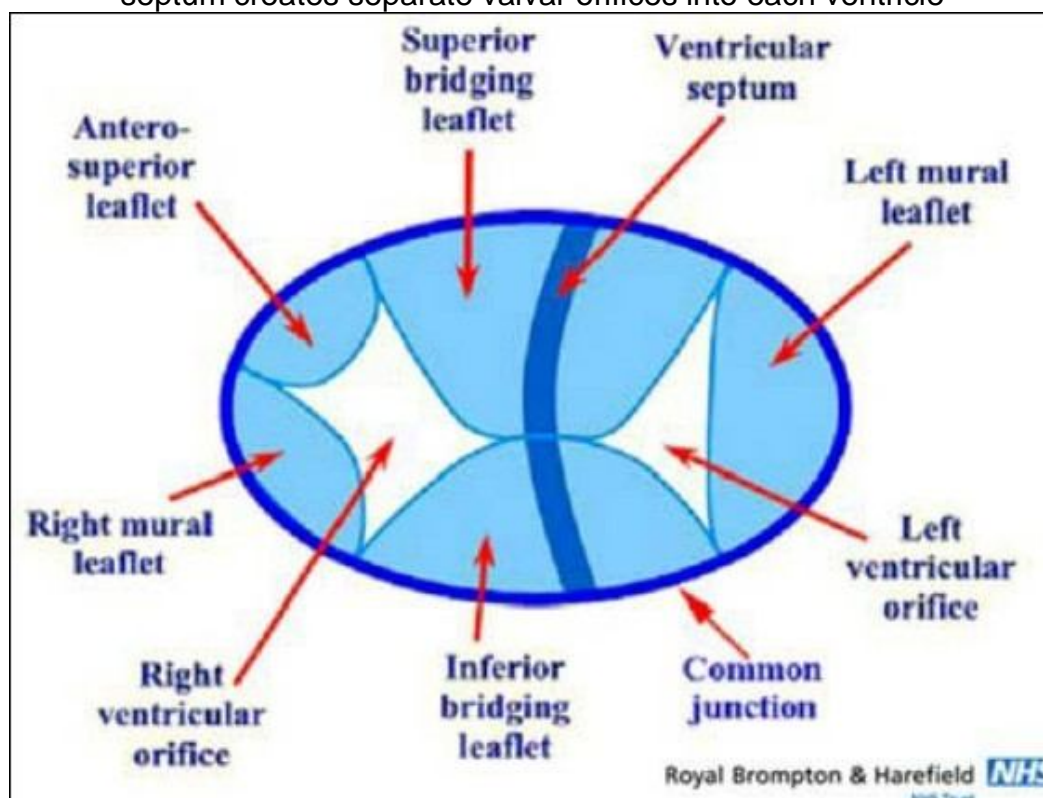
Figure 24 An ostium primum atrioventricular defect viewed from the base of the heart into the ventricular mass. It comprises a common atrioventricular junction, with separate valvar orifices to each ventricle, and an anteriorly displaced aorta



PV, pulmonary trunk. Ao, aorta

These anomalies, therefore, should be categorised as atrioventricular septal defects, even though shunting across the defect is exclusively at atrial level. The hearts do not have separate atrioventricular junctions as in the normal heart, and as in all other hearts described thus far with interatrial communications. In reality, the “ostium primum” defect, possesses all the phenotypic features of atrioventricular septal defect with common atrioventricular junction. The most important characteristic is that the valve guarding the common junction has five leaflets. Only two leaflets are exclusively within the right ventricle; and one is solely within the left ventricle. The remaining two leaflets, the superior and inferior bridging leaflets, are shared between both ventricles, straddling the ventricular septum (Figure 25).

Figure 25 Diagram representing the common atrioventricular junction in hearts with an ostium primum atrioventricular septal defect viewed from the base of the heart. The fusion of the superior and inferior leaflets across the ventricular septum creates separate valvar orifices into each ventricle



The distinguishing feature of the “primum” defect is that the two bridging leaflets are joined to each other by a tongue of fibrous tissue positioned directly on top of the crest of the ventricular septum, thus dividing the junction into discrete and separate left and right valvar inlets to the ventricles.<sup>30</sup> Almost always, the bridging leaflets and the tongue are also fused to the ventricular septal crest. It is this feature which confines shunting across the septal defect at atrial level, albeit with much of the shunting being below the level of the atrioventricular junction (Figure 26,27). The outflow tract of the left ventricle, nonetheless, retains the phenotypic anterior displacement, resulting in a

longer and narrower left ventricular outflow tract as the aorta is 'unfastened' from its central position at the base of the heart. Indeed, the tethering of the superior bridging leaflet to the ventricular septal crest elongates the length of the outflow tract when compared to hearts with common atrioventricular valvar orifice.

Figure 26 Long axis view of a heart with an ostium primum defect. The defect permits communication at atrial level

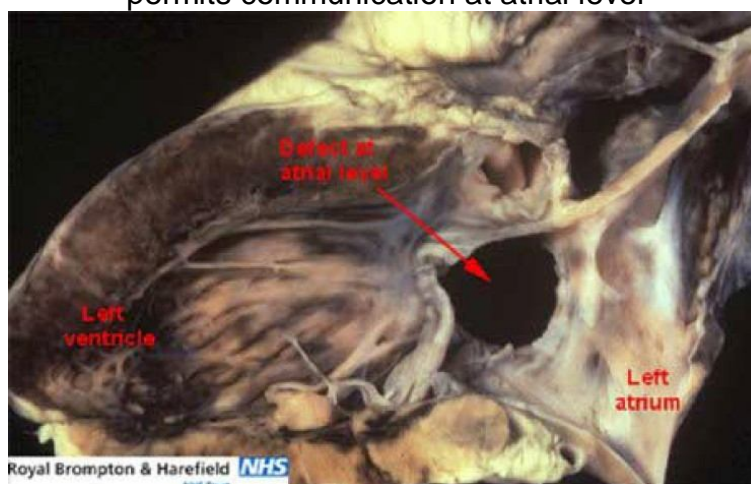
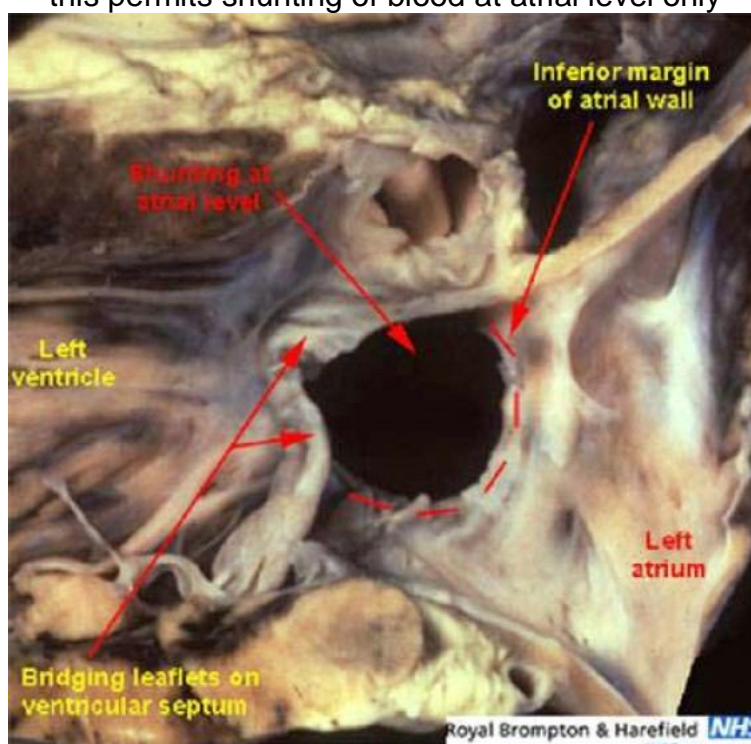


Figure 27 Long axis view of a heart with ostium primum defect. The inferior margin of the atrial septum does not connect with the ventricular mass, forming a common atrioventricular junction. Attachment of the left AV valve to the crest of the ventricular septum creates a separate orifice to each ventricle, this permits shunting of blood at atrial level only



### **Associated Considerations**

Atrial septal defects and interatrial communications can be accompanied by a variety of other congenital cardiac defects, such as coarctation of the aorta,<sup>31</sup> mitral valvar prolapse,<sup>32</sup> and partially anomalous drainage of the pulmonary veins into the right atrium.<sup>33</sup> Regardless of their anatomical location, the physiologic consequence of atrial septal defects and atrial communications result in the shunting of blood from one atrium to the other. Ultimately, the direction and magnitude of shunting is determined by the size of the defect, and the relative compliance of the ventricles. These vary according to age, with children having a normal compliance into infancy and a faster heart rate,<sup>34</sup> compared to adults who have a comparatively slower rate and a ventricular compliance that decreases with age.<sup>35</sup> Typically a sizable defect permits a large shunt, with significant hemodynamic consequences.

### **Conclusion**

The spectrum of interatrial communications is very broad, covering deficiencies of the flap valve of the oval fossa, to formation at various locations within the atrium of an extra-septal channel linking the right and left sides of the heart. Although generally grouped together as atrial septal defects, their phenotypic features are quite different and require careful distinction between defects that are located within the true atrial septum from those that are outside the confines of the oval fossa. Taken together, interatrial communications comprise one of the most common of all congenital cardiac defects, and often with an associated cardiac defect. The use today of advanced imaging techniques to assess the morphology and physiology of these frequent cardiac anomalies has led to efficient planning of treatment through surgical or interventional closure. These procedures now carry very low rates of morbidity and mortality, providing a positive outlook for the patient.

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