IMAGES in PAEDIATRIC CARDIOLOGY

Images Paediatr Cardiol. 2002 Jul-Sep; 4(3): 21–29.

PMCID: PMC3232521

Spontaneous closure of a large secundum atrial septal defect

N Galea and V Grech

Paediatric Department, St. Luke's Hospital, Guardamangia, Malta

Contact information: Dr. Victor Grech, Editor-in-Chief, Images Paediatr Cardiol, Paediatric Department, St. Luke's Hospital, Guardamangia - Malta ; Email: victor.e.grech@magnet.mt

Abstract

Large and untreated secundum atrial septal defects are closed in childhood in order to prevent significant rates of morbidity and mortality. Small defects often close spontaneously. We present a girl with a large atrial septal defect that underwent spontaneous closure at just over three years of age. The defect was haemodynamically significant and larger than conventional expectations for spontaneous closure.

MeSH: Atrial septal defect, Spontaneous resolution, Heart defects, congenital

Introduction

In foetal life, the flap valve of the foramen ovale hinges from the antero-superior rim of the fossa ovalis posterior to the aortic root. The rims of the fossa are composed of the infolded walls of the right and left atria, with the superior and posterior rims consisting of infoldings between the attachments of the caval veins to the right atrium, and the right pulmonary veins to the left atrium.

In the majority of individuals, the flap valve overlaps the fossa's rim, and at birth, when the left atrial pressure exceeds right atrial pressure, the flap valve is pushed against these rims, thus occluding the foramen.[1] Secundum atrial septal defects (ASD) are congenital deficiencies of the atrial septum in the fossa ovalis. Such defects are created when the flap valve is perforated or is too small to completely overlap the rim of the fossa.[1]

Large defects allow significant left to right shunting of blood, resulting in volume overload of the right heart and pulmonary vascular tree.[2] Untreated ASDs may cause a variety of complications. These include the eventual development of pulmonary hypertension, atrial arrhythmias and paradoxical embolisation with infarction of organs.[3]

ASDs may close spontaneously in childhood.[4] Persistent defects with pulmonary to systemic flow ratios (Qp/Qs) of >1.5 are operated before school age or whenever a diagnosis is made if later.[5] A variety of devices for transcatheter closure of ASDs have been developed and offer an alternative to surgical treatment.[6]

A previous study in Malta had found an incidence of ASD of 2.4/1000 live births.[7] These were divided into 2.0/1000 requiring intervention and 0.4/1000 live births not requiring intervention. 92% of defects not requiring intervention closed spontaneously, and the remainder had spontaneously decreased in size on follow-up.

We present a girl with a large atrial septal defect that underwent spontaneous closure.

Patient

Our patient (female) was referred by her general practitioner for a persistent systolic murmur at the age of two months. She was well, thriving and had no feeding problems or other manifestations of heart disease. This was a normal vaginal delivery at term, the first born child of healthy and unrelated parents.

Clinical examination showed no abnormalities other than a short 2/6 systolic murmur at the left sternal edge. Echocardiography at four months of age showed a large defect in the fossa ovalis of at least 11 mm diameter, with moderate right heart dilation. Slight infolding of the rim of the atrial septal defect was noted but the flap valve was not seen. Mitral valve inflow velocity (MVI) 0.7 m/s vs. tricuspid inflow velocity (TVI) of 1.15 m/s (figures 1 and 2). The clinical signs at this time were those of a significant atrial septal defect with a right ventricular heave, fixed splitting of the second heart sound and a loud pulmonary systolic murmur.

Figure 1 Subcostal 4 chamber view – secundum atrial septal defect

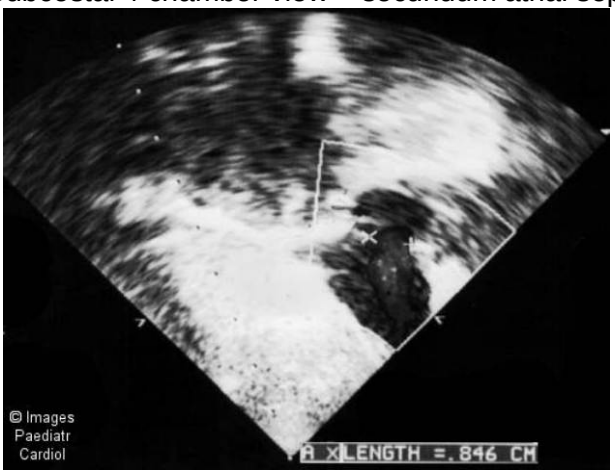
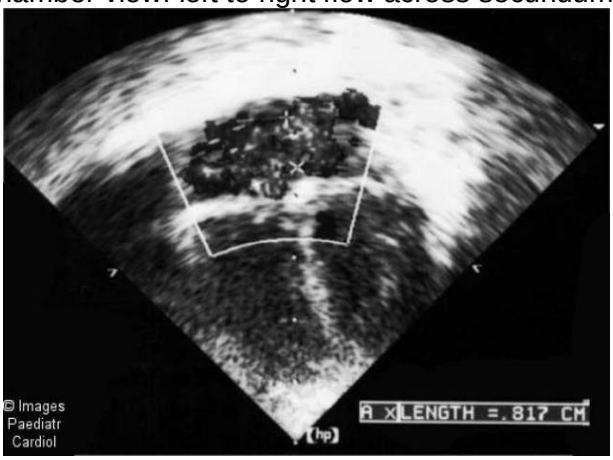


Figure 2 Apical 4 chamber view: left to right flow across secundum atrial septal defect



Repeat echocardiogram at seven months confirmed the diagnosis (MVI 0.8 m/s and TVI 1.2 m/s – figures 3 and 4). At this point, a perforated flap valve that of the foramen ovale was seen over the defect, with no obstruction to left to right shunting at this time. The valve appeared aneurysmal.

Figure 3 Flap valve formation across atrial septal defect

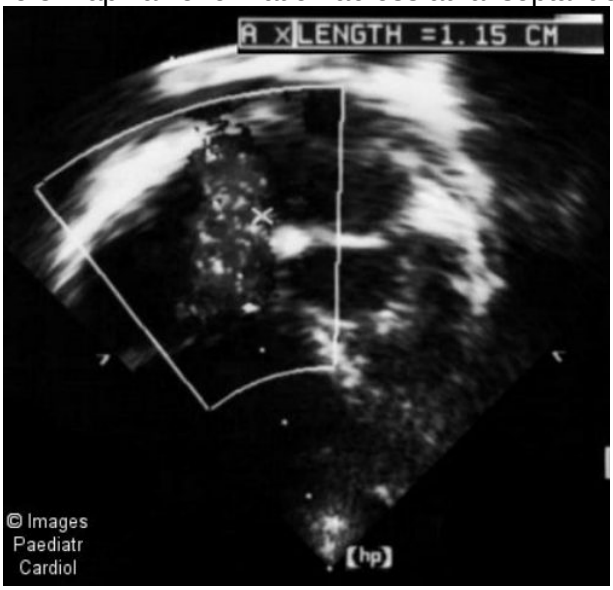
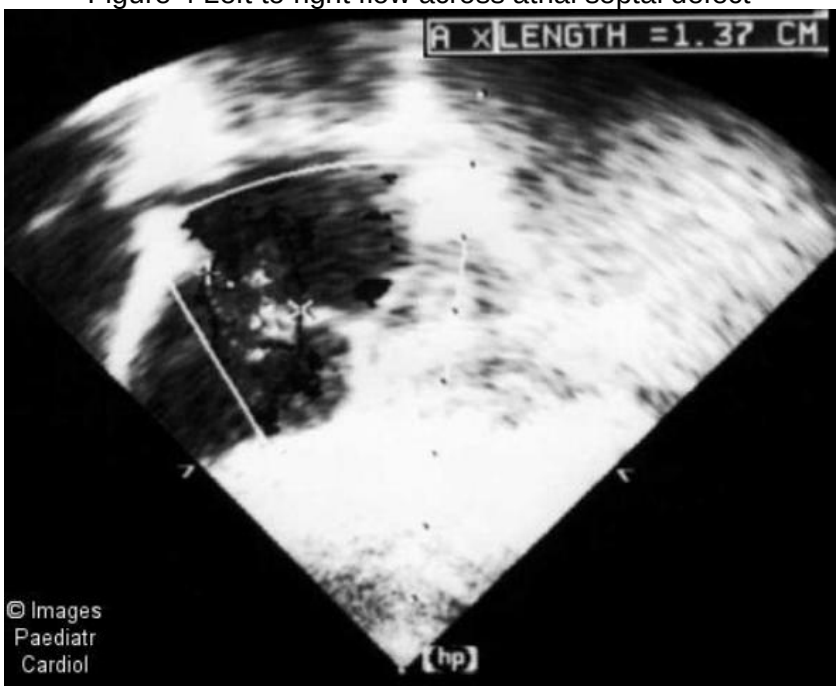
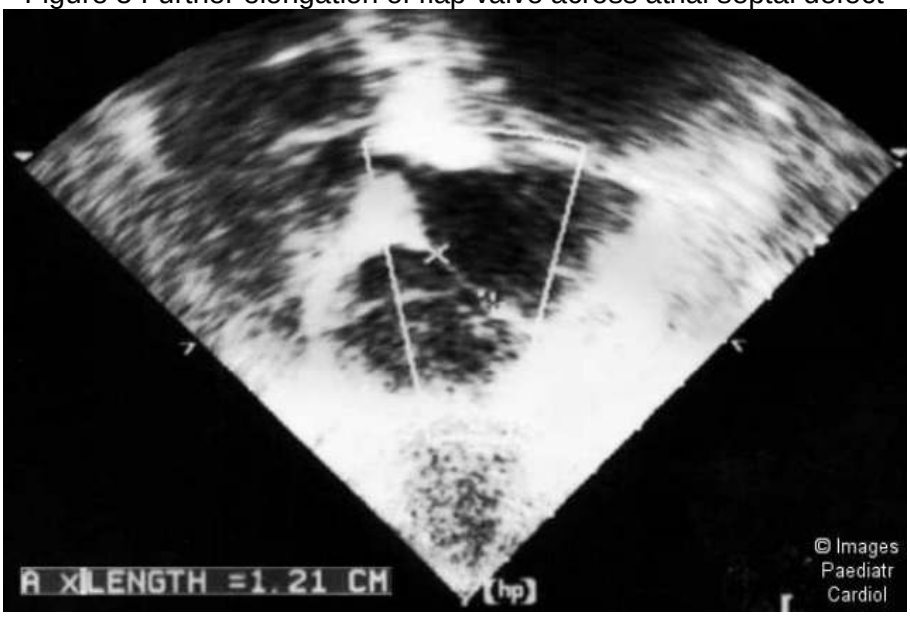


Figure 4 Left to right flow across atrial septal defect



Echocardiogram at nine months of age showed that the flap valve partially covered the defect, but there was still no obstruction to left to right flow with persistent right heart dilation (MVI 0.7 m/s and TVI 1 m/s – figure 5).

Figure 5 Further elongation of flap valve across atrial septal defect



An echocardiogram at 14 months of age showed that the defect was 5mm at its widest diameter with evidence of significant left to right shunting (MVI 0.6 m/s and TVI 0.9 m/s – figures 6 and 7). However, clinical examination at this time showed marked reduction in the intensity of the murmur and normal splitting of the second sound.

An echocardiogram at 23 months of age showed remarkable reduction in the size of the defect with no right heart dilation (MVI 1.1m/s and TVI 0.77 m/s – figs 8 and 9).

A final echocardiogram at of 3 years and 3 months of age confirmed complete resolution of the defect (figure 10) with completely normal echo and normal cardiovascular examination.

Figure 6 Residual defect – overall smaller

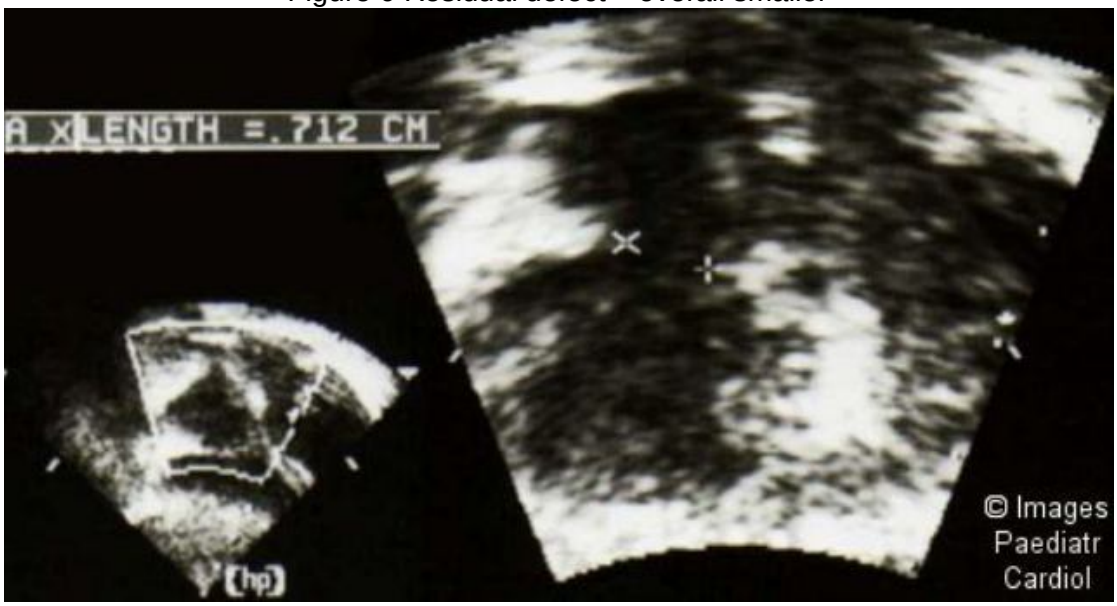


Figure 7 Residual defect – overall smaller

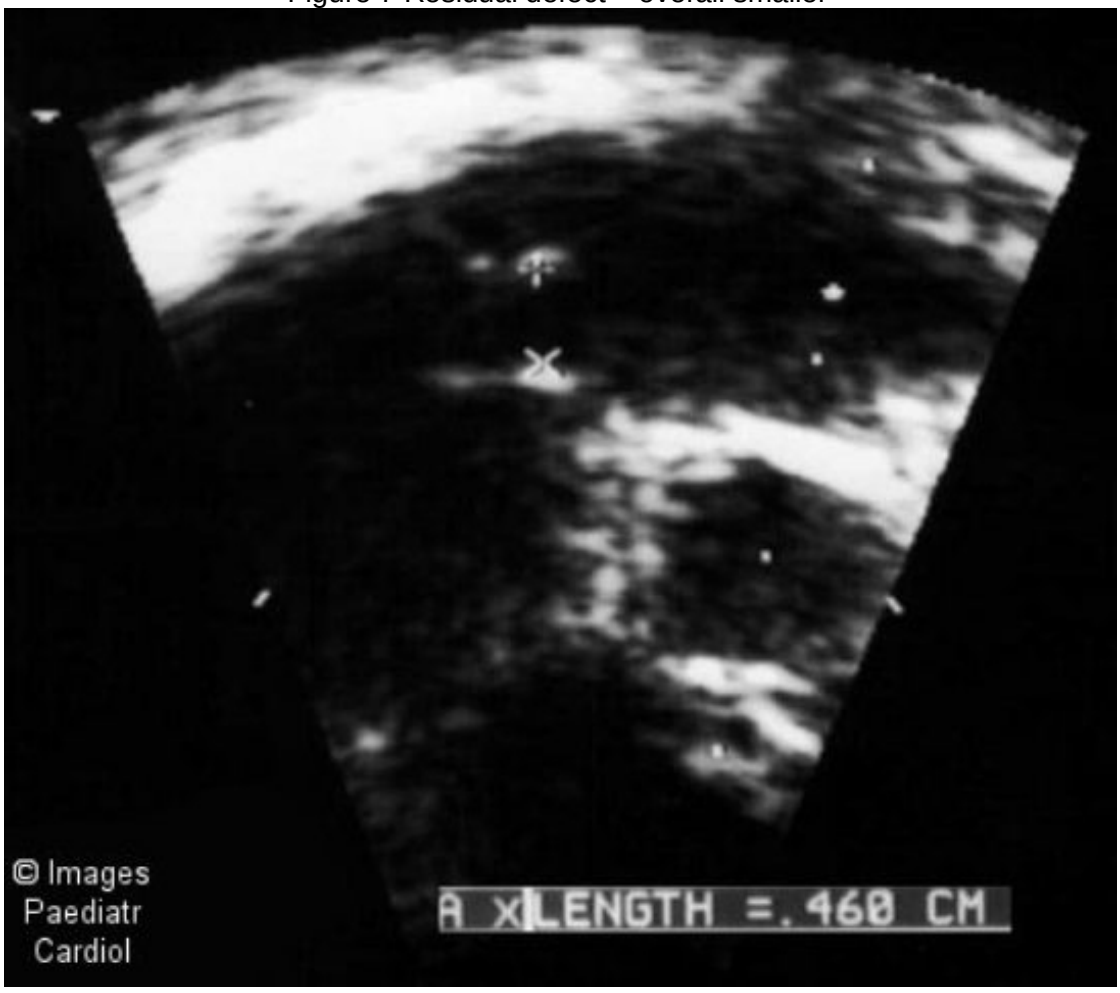


Figure 8 Further reduction in defect size

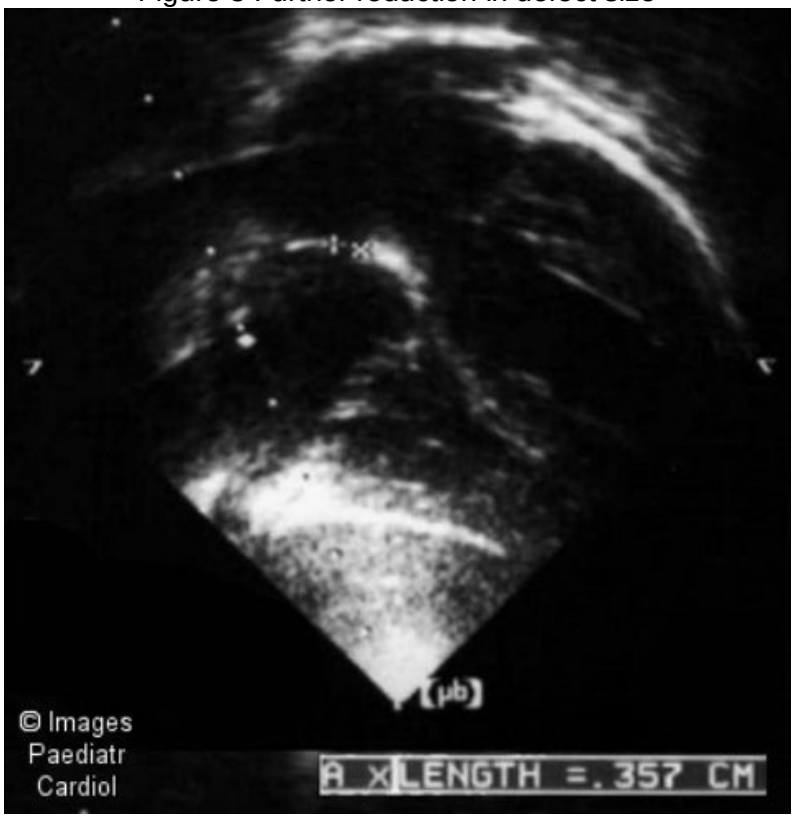


Figure 9 Further reduction in defect size

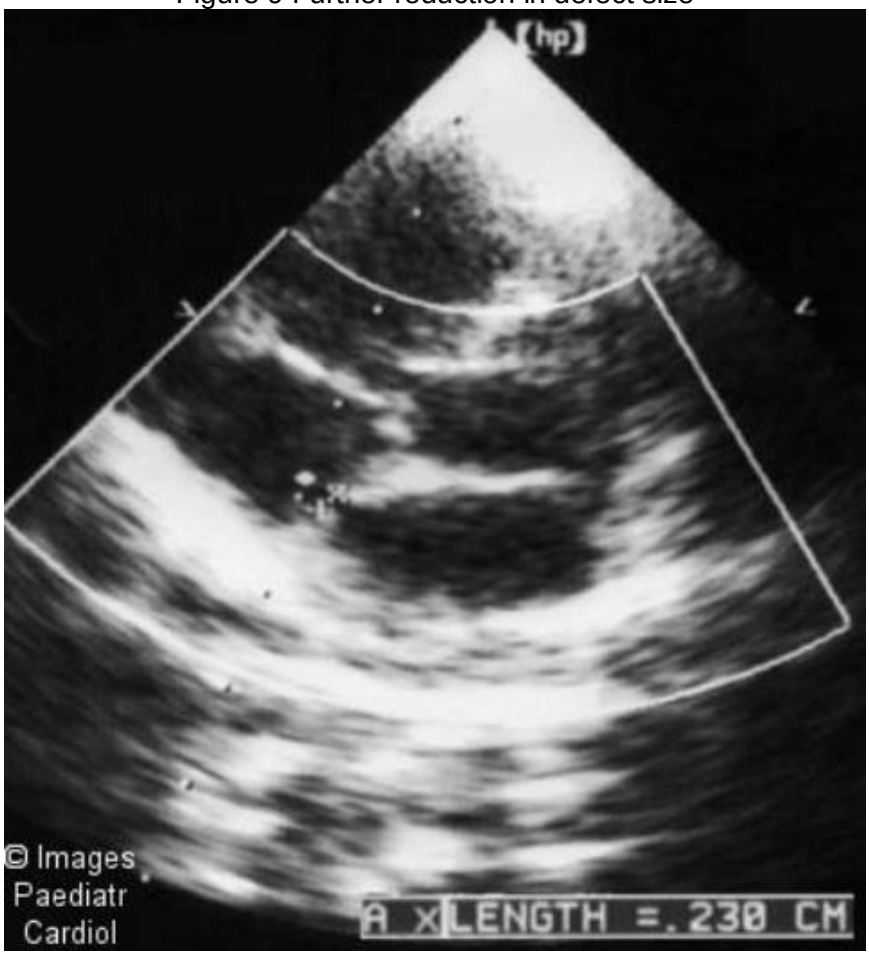
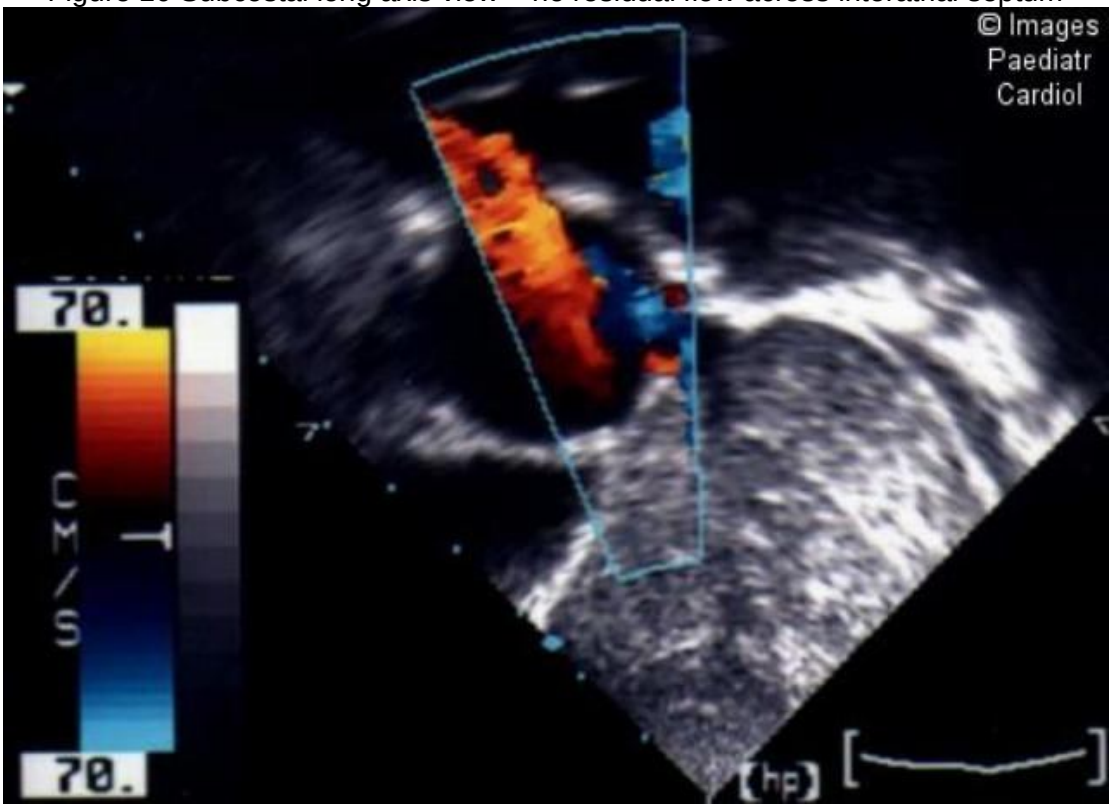


Figure 10 Subcostal long axis view – no residual flow across interatrial septum



Discussion

The first report of spontaneous closure of a small ASD documented by cardiac catheterisation was in 1966.[8] In the following year, spontaneous closure of defects which had been large and symptomatic was also reported.[9] Diagnosis after one year of age has been linked to a lesser likelihood of closure and this was found to be unrelated to the initial size at diagnosis.[10] Echocardiography confirmed that large defects causing right heart volume overload can close spontaneously.[11] Different processes of closure were demonstrated, including a valve mechanism at the fossa ovalis[12] and by aneurysm formation of the adjoining septum.[13] It is important to note that it is deficiency in the primary atrial septum that causes a defect in the fossa ovalis, and growth of this structure permits final spontaneous closure.

A prospective, echocardiographic study in 1993 on a series of infants diagnosed as having ASD by 3 months of age showed that the strongest predictive factor for closure was initial size at diagnosis. It was reported that ASDs with diameters ≥ 8 mm uniformly failed to close.[4] However, this finding was not borne out in a Maltese study as some defects with sizes at diagnosis in excess of 8 mm underwent spontaneous closure.[7]

Conclusion

Defects larger than 8mm with significant left to right shunting may undergo spontaneous closure, and in the absence of symptoms, our threshold for device closure should remain unchanged.

Acknowledgments

Mr. Iro Galea for scanning the images in this article.

References

1. Ferreira Martins JD, Anderson RH. The anatomy of interatrial communications--what does the interventionist need to know? Cardiol Young. 2000;10:464–473. [PubMed: 11049122]
2. Anderson RH, Macartney FJ, Shinebourne EA, Tynan M. Paediatric Cardiology. First Edition. Churchill Livingstone; 1987. pp. 541–560.
3. Craig RJ, Selzer A. Natural history and prognosis of atrial septal defect. Circulation. 1968;37:805–815. [PubMed: 5646864]
4. Radzik D, Davignon A, Van Doesberg N, Fournier A, Marchand T, Ducharme G. Predictive factors for spontaneous closure of atrial septal defects diagnosed in the first 3 months of life. J Am Coll Cardiol. 1993;22:851–853. [PubMed: 8354823]
5. Kirklin JW, Barratt-Bouyes G. Cardiac Surgery. Second Edition. Churchill Livingstone; 1993. pp. 609–644.
6. Ebeid MR. Percutaneous catheter closure of secundum atrial septal defects: a review. J Invasive Cardiol. 2002;14:25–31. [PubMed: 11773692]
7. Grech V. Atrial septal defect in Malta. J Paediatr Child Health. 1999;35:190–195. [PubMed: 10365359]
8. Timmis GC, Gordon S, Reed JO. Spontaneous closure of an atrial septal defect. J Am Med Assoc. 1966;196:137–139.
9. Cayler GG. Spontaneous functional closure of symptomatic atrial septal defects. New Eng J Med. 1967;276:65–70. [PubMed: 4224542]
10. Mody MR. Serial hemodynamic observations in secundum atrial septal defect with special reference to spontaneous closure. Amer J Cardiol. 1973;32:978–981. [PubMed: 4757238]
11. Ghisla RP, Hannon DW, Meyer RA, Kaplan S. Spontaneous closure of isolated secundum atrial septal defects in infants: an echocardiographic study. Am Heart J. 1985;109:1327–1333. [PubMed: 4003243]
12. Fukuzawa M, Fukushige J, Ueda K. Atrial septal defects in neonates with reference to spontaneous closure. Am Heart J. 1988;116:127–129.
13. Awan IH, Rice R, Moodie DS. Spontaneous closure of atrial septal defect with interatrial aneurysm formation. Pediatr Cardiol. 1982;3:143–145. [PubMed: 7155948]