

thrombus in the LAA might pose a danger to the systemic circulation (as usual) but not to the pulmonary given the highly restrictive tunnel-like connection with the RA. In an open-heart surgical procedure, it is possible to confuse the anatomy of the atrial septum but the size differentials between the other normal orifices and the tunnel entrance makes this improbable. The danger of this abnormality occurs during per-catheter creation of an atrial septal defect.

Balloon septostomy through the RATLAA, either by a moving balloon (Rashkind procedure) [3] or by balloon dilatation [2], could tear the atrial wall and result in exsanguination into the pericardium. This paper was written to help catheterizing cardiologists avoid this disaster.

### Technical Ways to Avoid Ballooning the RATLAA

- When the diagnostic catheter is positioned in the left atrium, record the image of catheter course for comparison when placing the BAS catheter (Fig. 1F).
- During balloon inflation, a properly positioned balloon bounces within the left atrium; when in the LAA, the balloon would be relatively immobile.
- The partly inflated balloon in the left atrium is smooth and round.

Through the RATLAA, the balloon may be asymmetric or even indented during the inflation process.

- When a properly positioned balloon is fully inflated, blood pressure drops as the balloon interferes with cardiac output. This should not occur with the balloon through the RATLAA.
- Danger! Given the distensibility of the LAA, inflating a balloon is unlikely to cause rupture. However, the tunnel is at risk either from the rapid pullback (Rashkind technique) or the lateral hoop stress of balloon dilatation.
- A balloon septostomy catheter with an end hole would be helpful by allowing injection of contrast agent.
- The definitive way to ensure proper position with current technology is by imaging with ultrasound. This can easily be done either in the catheterization laboratory (for those of us who still prefer to do balloon septostomy there) or in the neonatal intensive care unit.

### References

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## Around *PediHeart*: Noncompaction of the Left Ventricle

A *PediHeart* member from France presented a patient first diagnosed as a fetus and subsequently confirmed postnatally to have noncompaction of the left ventricular myocardium. Initially, there was mild to moderate left ventricular systolic dysfunction and he was begun on digoxin therapy. By one year of age the fractional shortening was normal. The baby had always been asymptomatic. The baby's doctor was concerned that the prognosis of this unusual pathology is not well known and there are risks from cardiac arrhythmias, thromboemboli, and cardiac failure. He wanted to get the group's suggestions on future management.

The responses were quite variable and ranged from those who would offer no pharmacological intervention at this point to several who recommended continuing the Digoxin, and adding an ACE inhibitor plus aspirin. All agreed that the natural history is highly variable [1]. The heart can become dilated or hypertrophic and systolic function can wax and wane. Restrictive cardiomyopathy and endocardiofibroelastosis have also been reported in association with a "spongy myocardium." Life-threatening arrhythmias can occur without warning and emboli can be a real problem with tragic results. I believe that, at a minimum, all respondents felt that close follow-up and frequent echocardiographic examinations were justified. Finally, this pathology has been found as part of several different syndromes, including the Roifman [2] and Melnick-Needles syndromes [3]. Two members also mentioned Barth Syndrome and would recommend screening the urine for organic acids.

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