The first case of transcatheter intervention in an adult with Cor Triatriatum Sinister

Breathing was difficult on 4 liters of oxygen. Her weight had slipped below 100 pounds. For 51 years there was a defect in her heart, a membrane that split her left upper heart chamber, the left atrium, into two chambers. This rare congenital (develops before birth) heart abnormality is known as Cor Triatriatum Sinister (CTS). The CTS membrane causes obstruction of oxygenated blood from the lungs entering the left atrium leading to increased pressure in the lungs. It is usually diagnosed in childhood, but a rare presentation is seen in adults when the membrane is incomplete allowing a small amount of blood to flow through it. Surgery is the first line of treatment in patients with CTS. However, when the patient presented at our clinic, she had two or more chronic diseases or conditions (comorbidities) and surgery was ruled out as a treatment option.

While resting, her oxygen saturation was 90% on 4 liters of oxygen delivered via a tube directly into her nose. Normal blood oxygen level is considered to be between 95-100% on room air. She was only able to walk 1218 feet during her six minute walk test whereas her predicted normal value was over 2000 feet. The physicians determined that a cardiac catheterization was necessary to treat her condition. Under sedation, the imaging modality of three-dimensional transesophageal echocardiography (3DTEE) was used to view and characterize the defect in her heart. The membrane had a 9 mm hole in it, which allowed a small amount of blood to flow through it (Fig. 1A).

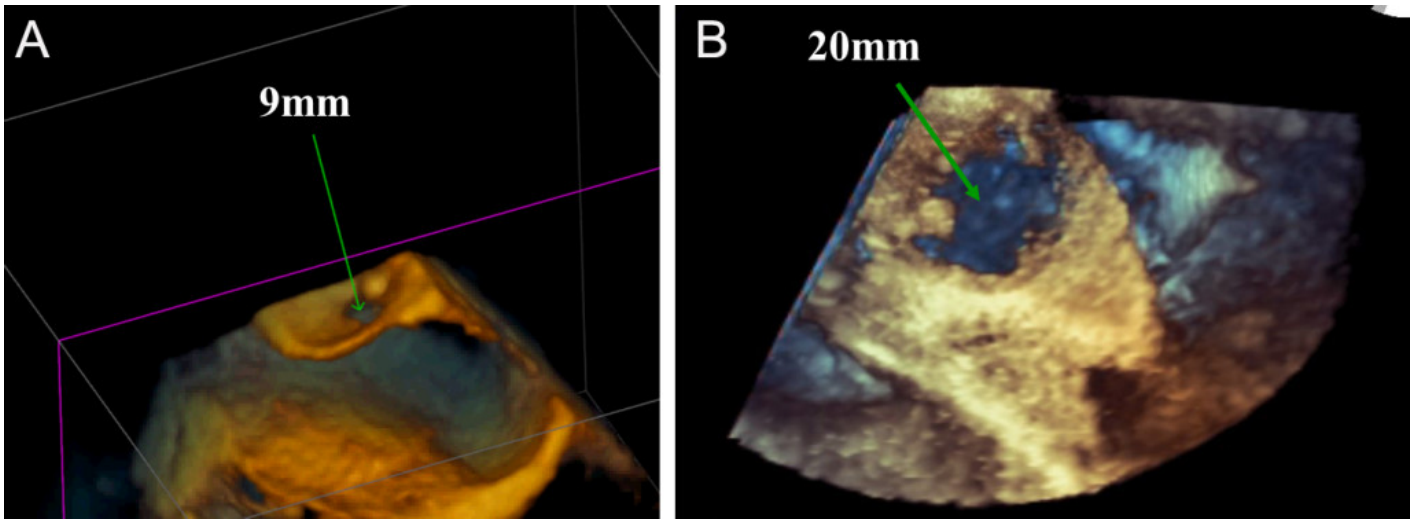

Fig. 1. A. 3DTEE imaging of the heart shows the CTS membrane that separated the left atrium into two chambers with a 9 mm hole that allowed restricted blood flow.
B. 3DTEE shows the CTS membrane after balloon dilation with a diameter of 20 mm.

A wire was inserted through the groin (right femoral vein) to the right side of the heart. A small puncture was then made from the right side of the heart into the left lower atrium (left distal atrium). A 20 mm balloon catheter was then inserted over the wire and positioned in the 9 mm hole in the membrane and inflated to dilate the membrane. The resulting 20 mm diameter of the CTS membrane allowed good blood flow decreasing the pressures in the left atrium and lungs (Fig. 1B). At her two-week follow-up visit, she had significant improvement in her symptoms and was able to perform daily activities with ease. Her six minute walk test distance significantly improved to 1,340 feet without the need for additional oxygen support.

Although the first line of treatment for CTS is surgery, our experience shows that transcatheter intervention may be considered in adults with multiple comorbidities.

Publication

Transcatheter intervention in cor triatriatum sinister.
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