Corrected transposition of the great vessels is a rare congenital cardiac malformation, in which the right ventricle supports the systemic circulation, whilst the left ventricle supports the pulmonary circulation. Systemic valve regurgitation can produce heart failure, as in the case described below.

Alfieri and colleagues reported the clover technique as an alternative method of correcting tricuspid regurgitation, similar to edge-to-edge mitral valve repair, with excellent mid-term results.\(^1,2\) We describe a unique case with management of significant tricuspid regurgitation using the edge-to-edge technique in an adult patient with corrected transposition of the great vessels, who presented with cardiac failure.

**Case presentation**

A 27-year-old Caucasian woman was admitted to our department for repair of tricuspid regurgitation in a congenitally corrected transposition of the great vessels with dextrocardia morphology. She presented with breathlessness, New York Heart Association (NYHA) class IV. She had been asymptomatic throughout her childhood. During her early twenties she became gradually symptomatic with exertional dyspnoea. She presented with symptoms and signs of heart failure.

Transoesophageal echocardiography revealed corrected transposition of the great vessels with a morphological left atrium draining into a right ventricle type chamber. The morphologically right ventricle supported the systemic circulation. The systolic function was satisfactory. The morphological right atrium drained into a morphological left ventricle, which supported the pulmonary circulation. The tricuspid valve (systemic atrioventricular valve) was grossly incompetent, with apical displacement in keeping with Ebstein's anomaly. The interatrial septum was intact, with evidence of a small patent foramen ovale. Despite optimal medical therapy, the patient’s symptoms worsened and she was referred for surgery.

The patient was placed on cardiopulmonary bypass. The systemic atrioventricular valve was approached via a superior transseptal approach. Access was made extremely difficult by the anticlockwise rotation of the heart. The right ventricle was dilated. Inspection of the systemic atrioventricular valve showed it to have a huge di-
ameter — approximately 100 mm — and a grossly distorted anatomy. There were two orifices. The first was a defect on what looked like a posterior leaflet, with no underlying support. The second was the main valve orifice, which was grossly incompetent.

The defect in the posterior leaflet was closed in double layer fashion using continuous 5(0) polypropylene suture buttressed with pericardium. The major valve orifice was approximated in its middle using Alfieri stitch. It was not technically feasible to insert a ring because of the large valve circumference and the complete absence of an underlying valve ring to permit the deployment of strong sutures to hold. The patient was weaned off cardiopulmonary bypass without difficulty. She made an uneventful postoperative recovery, and was discharged home 6 days later.

Postoperatively, the patient improved significantly. Three years post-procedure, her NYHA status is grade I. On repeat transthoracic echocardiography, the repaired tricuspid valve was noted to have Ebstein’s anomaly as before, with mild regurgitation (Figure 1a and b).

Discussion
Corrected transposition of the great vessels is a rare congenital cardiac defect, involving left-handed looping of the heart tube, resulting in atrioventricular (AV) discordance, while the aorto-pulmonary septum fails to rotate through 180°, resulting in ventriculo-arterial discordance. Essentially the vessels are transposed, but blood flows in an effective sequence, hence the name corrected: the right ventricle supports the systemic circulation, with the left ventricle supporting the pulmonary circulation. Tricuspid regurgitation is one of the consequent pathologies and can result in heart failure.

Ebstein’s anomaly, first described by Wilhelm Ebstein in 1866, is characterised by apical displacement of the septal and posterior tricuspid valve leaflets, leading to atrialisation of the right ventricle with a variable degree of malformation and displacement of the anterior leaflet. Surgery is only indicated if the patient is haemodynamically compromised. Valve replacement was the treatment of choice until the mid 1970s, but was associated with high mortality and morbidity. Valve repair is now the procedure of choice.

We decided to perform the edge-to-edge technique to repair the valve because of the existing particularities: the huge size of the valve, the leaflet abnormality and the absence of any anatomical annulus. Therefore, it was impossible to perform annuloplasty. Moreover, as the diameter was about 100 mm, we could not replace it. The combination of defect repair and the edge-to-edge repair offered the desirable result in a difficult anatomical setting. Although it is the first report of edge-to-edge repair in congenital cardiac pathology, the combination of edge-to-edge repair with bicuspidalisation has previously been described as an effective method for treating acquired tricuspid regurgitation, while edge-to-edge repair has also been applied for the treatment of traumatic tricuspid regurgitation. Evidence now suggests that Alfieri repair with an annuloplasty ring, in the case of mitral regurgitation, gives good long-term results. We could not insert a ring in this situation because of the anatomical situation, as described above. This combination of edge-to-edge technique with bicuspidisation can provide at least an alternative useful medium-term repair in patients with a distorted anatomy who are unsuitable for annuloplasty.
References


