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CLINICAL SPOTLIGHT

# Headache in a Patient with Complex Congenital Heart Disease: Diagnostic and Therapeutic Considerations

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Headache in adult patients with congenital heart disease may be a manifestation of the underlying cardiac condition or more common alternative causes of headache. In patients with pre-existing congenital heart disease, consideration of potentially uncommon aetiologies of headache is important.
We report an uncommon case of headache in a patient with complex congenital heart disease characterised by Ebstein's anomaly with previous surgical repair complicated by idiopathic intracranial hypertension. This case illustrates the importance of understanding the implications of headache with reference to the underlying cardiac disease as well as specific issues related to a relatively young cohort of patients.

**Keywords** Ebstein's anomaly • Idiopathic intracranial hypertension • Glenn shunt • Tricuspid regurgitation

Headache in adult patients with congenital heart disease may complicate their specific underlying cardiac condition or be a result of the myriad of more common alternative causes of headache. In patients with headache and pre-existing congenital heart disease, consideration of potentially uncommon aetiologies is important, such as hypertension in a patient with previous aortic coarctation repair, cerebral abscess in a patient with Eisenmenger's syndrome or the reported association between migraine and intracardiac shunt.

We report an uncommon cause of headache in a patient with complex congenital heart disease, in which a 23-year-old woman with a background of Ebstein's anomaly and previous bidirectional Glenn shunt developed idiopathic intracranial hypertension. This case illustrates the importance of understanding the implications of headache with reference to the underlying cardiac disease as well as specific issues related to a relatively young cohort of patients.

The patient was diagnosed with Ebstein's anomaly and atrial septal defect in infancy and was initially palliated with a right Blalock shunt. At the age of five years, she subsequently underwent tricuspid valve repair using Carpentier technique with plication of the right atrium and ventricle with suture closure of the atrial septal defect. Due to ongoing effort intolerance with associated severe tricuspid incompetence, right atrial and right ventricular dilatation, further surgery was undertaken at age 19. Diagnostic cardiac catheterisation prior to surgery demonstrated a low transpulmonary gradient and low pulmonary vascular resistance (mean pulmonary artery pressure 9 mmHg, mean right atrial pressure 6 mmHg, left ventricular end diastolic pressure 6 mmHg, transpulmonary gradient 3 mmHg, pulmonary vascular resistance 0.8 Woods units). The patient then underwent tricuspid valve repair, tricuspid valve annuloplasty (31 mm Duran band), right atrial reduction and bidirectional superior cavopulmonary (Glenn) shunt creation. The patient remained limited by exertional fatigue and dyspnoea following her last corrective procedure, which were attributed to severe residual tricuspid regurgitation (Figure 1). Cardiopulmonary exercise testing confirmed exertional hypoxaemia, with pulmonary embolism and pulmonary AV malformation subsequently excluded. These symptoms were effectively managed by diuretic therapy. The patient's cardiac status

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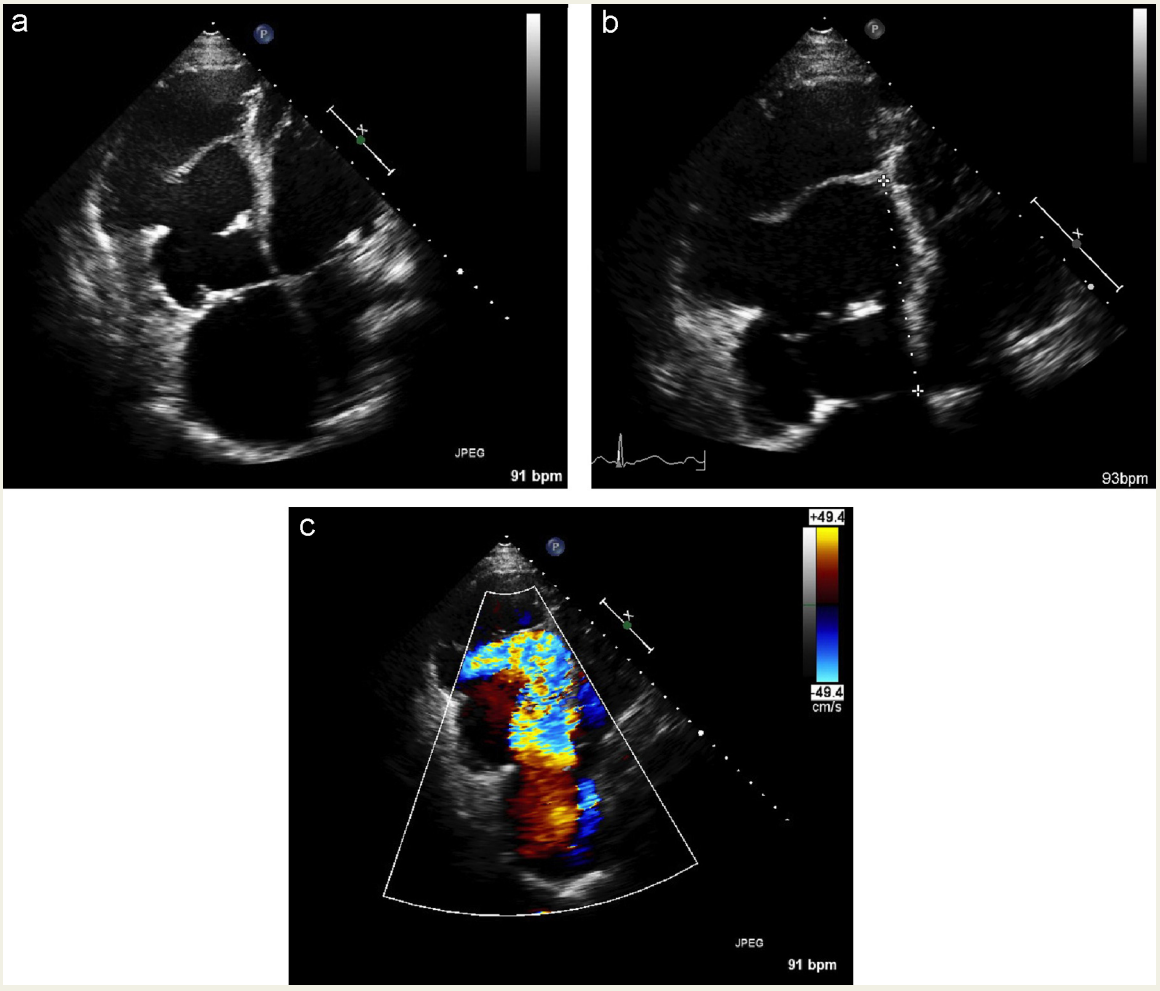


**Figure 1** Apical four-chamber images demonstrating severe apical tricuspid valve displacement consistent with known Ebstein's anomaly (Figures 1a and 1b). There is severe tricuspid valve incompetence as noted in Figure 1c.

subsequently remained stable apart from the development of atrial arrhythmia, which was managed by successful catheter ablation. The patient was maintained on no regular medical therapy aside from contraception in the form of Implanon® (etonogestrel), a progesterone-based implantable contraceptive, which delivered subcutaneously provides up to three years of contraception.

The patient then presented to emergency department with a two-week history of headache, visual disturbance and tinnitus. The patient denied diplopia. Clinical examination was unremarkable for infective or inflammatory signs. Visual acuity was reduced bilaterally. Visual fields were restricted and blind spots were enlarged. Fundoscopy revealed marked oedema of the right optic disc and mild oedema of the left disc. The remainder of her cranial nerves and limb examination were normal.

Lumbar puncture revealed raised cerebrospinal fluid (CSF) pressure at 33.5 cm. Lab examination of the CSF fluid showed normal cells and protein count.

Magnetic resonance imaging of the brain (Figure 2) showed cupping of the optic discs and enlargement of surrounding CSF with flattening of pituitary gland consistent with raised intracranial pressure. Magnetic resonance venography revealed bilateral transverse sinus stenosis. Echocardiography again demonstrated diminutive right ventricular size with severe tricuspid incompetence. Flow within the superior vena cava was phasic with antegrade flow noted from the right ventricle into the main pulmonary artery; there had been no interval change in the patient's echocardiographic appearance.

Following subsequent formal neurology review, the patient was commenced on acetazolamide 250 mg three times a day. Following three weeks of therapy, the patient noted a dramatic symptomatic improvement with improved results on visual field testing. Management was complicated, however, by medication side-effects including nausea, vomiting and paraesthesia. Due to problematic side-effects, the acetazolamide dose was halved with return of