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Images in Congenital Cardiac Disease

A classical case of the Gasul phenomenon

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Abstract This case demonstrates the development of secondary infundibular stenosis in a 10-year-old male child with documented large non-restrictive perimembranous ventricular septal defect in infancy – the classical Gasul phenomenon.

Keywords: Ventricular septal defect; Gasul phenomenon; infundibular stenosis

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A 10-year-old boy presented with insidious effort intolerance and poor growth. Past medical records documented a large perimembranous ventricular septal defect diagnosed at 4 months of age, with symptoms of heart failure (Fig 1a, parasternal short-axis view; Supplementary video 1). Advice for early surgical closure was not followed; notwithstanding, there had been gradual symptomatic improvement after infancy. Further examination revealed oxygen saturation of 98%, a normal-sized heart, and a harsh grade 4 ejection systolic murmur best appreciated in the left parasternal region. There was no associated click. Chest radiography showed a cardiothoracic ratio of 0.55 and right ventricular type of cardiac apex (Supplementary figure 1). Sinus rhythm with prominent right ventricular forces was observed in the 12-lead electrocardiogram (Supplementary figure 2). Echocardiography with colour flow imaging demonstrated a large 10-mm ventricular septal defect with the left-to-right shunt effectively restricted by a hypertrophic band in the right ventricular outflow tract (Fig 1b [arrow], parasternal short-axis view; Supplementary video 2) with peak systolic gradient of 84 mmHg (Fig 1c, continuous wave Doppler); biventricular function was normal, and cardiac catheterisation was confirmatory (Fig 1d [arrow], lateral projection; Supplementary figure 3, video 3). It showed elevated right atrial a-wave and proximal right ventricular systolic pressure with normal outflow tract and pulmonary artery pressures (Supplementary figures 4–6). Surgical correction was performed with patch closure of the septal defect and resection of the hypertrophic fibromuscular band (Fig 1e [arrow]; PV, pulmonic valve). Recovery was uneventful, and the child did well thereafter (Fig 1f, post-operative echocardiogram in the short-axis view showing intact septum and no residual infundibular stenosis).

Benjamin Gasul, in 1957, was the first to describe the intriguing phenomenon of secondary infundibular stenosis in ventricular septal defect.[1] Large case series on natural history in the following two decades found this occurrence in 3–7% of defects.[2,3] Hypertrophy of the crista supraventricularis in response to the stress of torrential pulmonary blood flow and elevated pulmonary arterial pressure is the mechanism postulated for this "acquired Fallot's tetralogy".[4] Contributory anatomical factors include a large subaortic defect, oblique infundibular angle (40–60°), >30% aortic override, anomalous right ventricular muscle bundles, and right aortic arch; it is one of the causes of amelioration of heart failure symptoms in an infant with a large ventricular septal defect.

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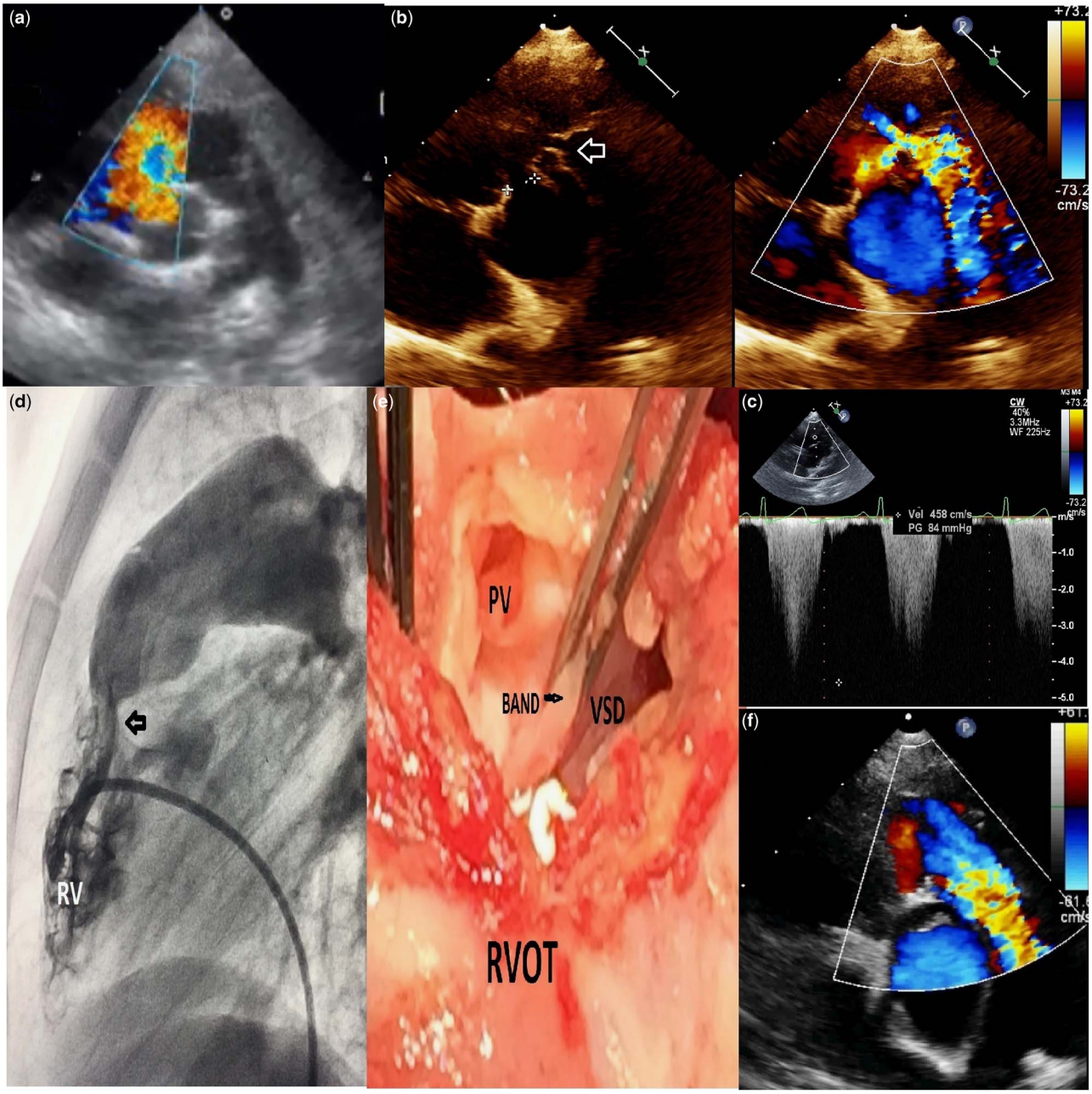

Figure 1.
Echocardiographic and angiographic demonstration of acquired infundibular stenosis in VSD, with surgical images. PV = pulmonic valve; RV = right ventricular; RVOT = right ventricular outflow tract; VSD = ventricular septal defect.

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Conflicts of Interest

None.

Supplementary material

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References

1. Gasul BM, Dillon RF, Vrla V, Hait G. Ventricular septal defects; their natural transformation into the cyanotic or noncyanotic type of tetralogy of Fallot. J Am Med Assoc 1957; 164: 847–853.
2. Weidman WH, Blount SG Jr, DuShane JW, et al. Clinical course in ventricular septal defect. Circulation 1977; 56 (Suppl): I56–I69.
3. Corone P, Doyon F, Gaudeau S, et al. Natural history of ventricular septal defect. A study involving 790 cases. Circulation 1977; 55: 908–915.
4. Watson H, Lowe KG. Functional adaptations of the right ventricular outflow tract in congenital heart disease. Br Heart J 1965; 27: 408–412.