Challenging Combination; Pseudocoarctation with Subaortic and Subpulmonic Stenosis

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Abstract
Combined left and right ventricular outflow tract obstruction is an extremely rare congenital anomaly. Pseudocoarctation (PcoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta. A 4-year-old girl presented to our hospital with progressive complaints of exertional dyspnea and intermittent dizziness for about one year. She was diagnosed with severe combined left and right ventricular outflow tract obstructions associated with pseudocoarctation of the aorta. Echocardiographic estimated peak pressure gradients across the right and left ventricular outflow tracts were 151 mmHg and 109 mmHg respectively. There was no significant pressure gradient across the pseudocoarctation. Aggressive surgical myomectomy for both outflow tracts, combined with reconstruction of right ventricular outflow tract using pericardial patch and iatrogenic ventricular septal defect (VSD) closure using PTFE patch was performed. No intervention was done for pseudocoarctation of the aorta (PcoA). Association of PcoA with different types of cardiac anomalies has been reported. However to our knowledge, combined left and right ventricular outflow obstruction associated with PcoA has not been described before.

Keywords
Pseudocoarctation; Aortic Stenosis; Critical Pulmonary Stenosis
Introduction
Although left and right ventricular outflow tract obstructions are among the most common congenital heart defects, the combination of both in the same patient appears to be very uncommon[1]. Pseudocoartation (PcoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta[2]. We describe a successful surgical treatment of congenital combined left and right ventricular obstruction associated with pseudocoarctation of the aorta in a 4 year old girl.

Case Report
An 4-year-old female was admitted to the cardiovascular department with one year history of exertional dyspnea, dizziness and intermittent confusion. Her family and past medical history was unremarkable. On physical examination, she was in good condition with a blood pressure of 90/60 mmHg, heart rate of 100-110 beats/min, respiratory rate of 21/min, and body temperature of 36 °C. Palpation of the chest revealed a thrill and on auscultation there was a grade 5/6 harsh, systolic ejection murmur associated with a mild diastolic murmur best heard along the left sternal border. Basic laboratory tests and cardiac enzyme levels were normal. The chest X-ray showed an increased cardiothoracic ratio and diminished pulmonary vascular markings. Electrocardiography showed sinus rhythm with biventricular hypertrophy and strain pattern. Echocardiography revealed severe hypertrophy of the left ventricle posterior wall (LV), right ventricle (RV) free wall, and interventricular septum (IVS). There were apparent muscular bundles at the level of the RVOT causing low-lying type stenosis. Estimated maximal pressure gradients across the right and left ventricular outflow tracts (LVOT) were 151 mmHg and 109 mmHg respectively (Figure 1). There was mild aortic regurgitation. Angiography demonstrated severely hypertrophied right and left ventricles, severe sub-valvular aortic and pulmonary stenosis (Figure 2). Pressure measurements revealed a peak-to-peak systolic gradient of 95 mmHg at LVOT and 140 mmHg at RVOT. In addition aortic an-
and valvular pulmonary stenosis, sub-valvular, and valvular aortic stenosis and hypertrophic cardiomyopathy, who presented with pulmonary hemorrhage[1]. Pseudocoarctation (PCoA), also known as kinking or buckling of the aorta, is an uncommon anomaly of the aorta[2]. In our patient LVOT obstruction was mainly sub-valvular, aortic annulus size was acceptable and the aortic valvular insufficiency was mild. For this reason we performed a very aggressive circumferential myomectomy to relieve the obstruction. Occurrence of third-degree complete A-V block and iatrogenic VSD undoubtedly is the result of very aggressive surgical approach in order to remove the LVOT obstruction. An aggressive circumferential myomectomy was performed. Valeske et al. reported that AV block was most likely related to the type of LVOT obstruction. In short discrete lesions the incidence of permanent complete AV block was none. However in complex LVOT obstruction like in our patient the incidence was up to 23%[2]. A variety of abnormalities were reported in association with pseudocoarctation, including bicuspid aortic valve, aortic stenosis, ventricular septal defect, atrial septal defect, patent ductus arteriosus[3]. PCoA is different from true coarctation in the following aspects (a) Unlike true coarctation, in which the aortic arch does not reach clavicle, the aortic arch of pseudocoarctation is elongated (b) Absence or mild degree of stenosis of aortic lumen. (c) No or small blood pressure gradient across the lesion (Δp< 25 mmHg). (d) Absence of collateral circulation[4]. PCoA used to be considered a benign lesion requiring no treatment. Steinberg et al. reported that congenital aortic stenosis and pseudocoarctation with four cases in autopsy series[5]. In our patient hemodynamic data and angiography along with physical examination correlated well with these rules. Since the pressure gradient across the PCoA was 10 mmHg and the patient was normotensive, no intervention was needed. Many reports showed that combination of PCoA with cardiac anomalies but to our knowledge, combined LVOT and RVOT obstruction with PCoA has not been described before.

Competing interests
The authors declare that they have no competing interests.

References