Fetal and neonatal percutaneous aortic balloon valvuloplasty in critical aortic stenosis followed by complex Ross–Rastan–Konno reconstruction

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A midgestation male fetus (22nd week of pregnancy) was diagnosed with critical aortic stenosis (AS) and concomitant left ventricular hypoplasia characteristic of progressive hypoplastic left heart syndrome (HLHS). To prevent heart dysfunction over the subsequent course of gestation and to reverse the evolution into HLHS, an intrauterine aortic balloon valvuloplasty was performed in the 26th week of pregnancy in Pediatric Heart Center (Linz, Austria). The size of the balloon was 4 mm, decompressed to 4.4 mm at the pressure of 16 bar. No complications occurred, and the procedure resulted in an increase of fetal left ventricular ejection fraction with flow normalization both in the ductus arteriosus and foramen ovale as well as resolution of mitral regurgitation. Owing to a recurrent constriction of the aortic ostium, an urgent percutaneous balloon valvuloplasty was performed immediately after birth (FIGURE 1A and 1B). Furthermore, the procedure was repeated after 15 months due to recurrence of AS. At the age of 5 years, control catheterization confirmed a stable course of hemodynamically significant complex stenosis of the left ventricular outflow tract (LVOT) with fibroelastosis and aortic regurgitation (FIGURE 1C). Concomitant losses of consciousness were a clear indication for an urgent surgical treatment.

A Ross–Rastan–Konno procedure was the final step in the treatment of the 7-year-old patient after the previous interventions during the fetal and neonatal periods. After a meticulous resection of the LVOT, endocardial fibroelastosis, and muscular hyperplasia, the top section of the resected interventricular septum was supplemented with a pulmonary autograft and an extensive fragment of the right ventricular free wall was implanted into the subaortic position. The operation included a vast reconstruction of the right ventricular outflow tract with the use of a flared pulmonary xenograft. The postoperative course was uncomplicated (FIGURE 1D). The patient was referred to an outpatient clinic, and his condition in the mid-term follow-up was good.

Fetal aortic balloon valvuloplasty for congenital AS may preserve the function of the left heart, thus preventing fatal intrauterine progression into HLHS.[1] Percutaneous and surgical interventions in newborns should be a spontaneous continuation to achieve biventricular outcome. With an optimal management of complex LVOT obstruction, patients could be referred for the final modified Ross–Rastan–Konno procedure for left ventricular function preservation, which is a feasible option in this population.[1,2]

The management of HLHS is a complex, multistaged (a minimum of 3 surgical procedures), and long-term univentricular palliation. The mortality rate exceeds 90% if the condition is left untreated.[3] Fetal cardiac interventions for congenital heart defects such as AS with HLHS physiology may prevent progression to complete HLHS, thus improving fetal survival, or may even help achieve biventricular outcome, as in our patient. On the other hand, fetal aortic valvuloplasty still carries a 30% risk of impaired pregnancy, including fetal demise.[1,4] Precise anatomical predictors of a successful biventricular outcome are still uncertain, and the results may vary among fetuses with critical AS or depending on institutional

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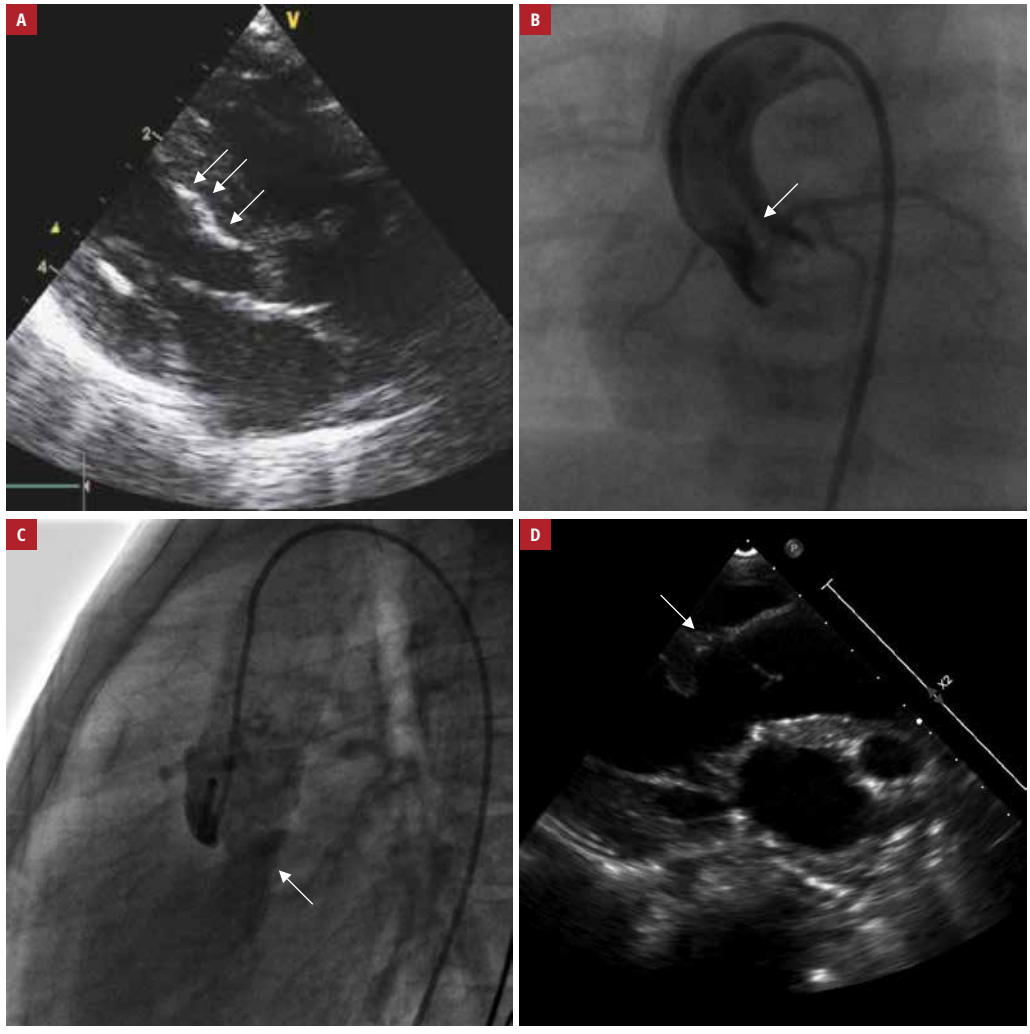

FIGURE 1 **A** – neonatal echocardiography: complex stenosis of the left ventricular outflow tract (LVOT), fibroelastosis in the LVOT (arrows), and dysplastic aortic valve after fetal balloon valvuloplasty; **B** – interventional neonatal balloon valvuloplasty for LVOT obstruction: the level of the aortic valve (arrow); **C** – follow-up catheterization: complex stenosis of the LVOT with progressive aortic valve regurgitation (arrow); **D** – follow-up echocardiography after the Ross–Rastan–Konno procedure: a resected interventricular septum supplemented with the pulmonary autograft and an extensive fragment of the right ventricular free wall implanted into the subaortic position (arrow)

experience.[4] Nevertheless, fetal aortic valvuloplasty provides a chance for a left heart rescue and further therapy, including cardiac surgery such as the modified Ross–Rastan–Konno procedure, to improve the patient's long-term prognosis.

In summary, an intrauterine cardiac intervention followed by neonatal aortic valvuloplasty preserved left heart function and allowed subsequent successful management with surgical LVOT reconstruction using the modified Ross–Rastan–Konno procedure.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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