Cardiology in the Young

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Early dehiscence of a tricuspid valve annuloplasty ring in an adolescent with hypoplastic left heart syndrome presenting with unconjugated hyperbilirubinemia

David Kielmayer[1], Eva Sames-Dolzer[2,3] and Gerald Tulzer[1,3]

[1]Department of Pediatric Cardiology, Kepler University Hospital, Linz, Austria; [2]Division of Pediatric and Congenital Heart Surgery, Kepler University Hospital, Linz, Austria and [3]Medical Faculty, Johannes Kepler University Linz, Linz, Austria

Brief Report

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Corresponding author:
David Kielmayer;
Email: david.kielmayer@kepleruniklinikum.at

Abstract

We report a unique case of an adolescent patient with Fontan physiology presenting with unconjugated hyperbilirubinemia due to dehiscence of a tricuspid valve annuloplasty ring.

Case report

A 14-year-old adolescent of Kyrgyz origin with hypoplastic left heart syndrome and Fontan circulation and an implanted dual chamber (DDD) pacemaker—sought medical attention in the emergency department due to jaundice for 10 days. He was discharged under best conditions from hospital a month earlier after tricuspidal valve repair via DeVega annuloplasty with a 30 mm Edwards® MC3 Tricuspid annuloplasty ring. On admission, discrete dermal and scleral icterus, a 3/6 systolic heart murmur, and an arrhythmical heart rhythm were found. There was no history of discoloured stool or reddish/brown urine and pruritus was denied. Furthermore, there was no history of fever, oedema, decreased physical capacity, increasing cyanosis, or respiratory distress. Blood pressure was 94/54 mmHg (right upper extremity), heart rate was 84 beats per minute, and saturation values averaged at 87%.

The patient was admitted to the Department of Pediatric Gastroenterology and Hepatology for further investigation where laboratory evaluation showed haemolysis without anaemia (haemoglobin: 13.1 g/dL) with a high reticulocyte count, reflecting an increased erythropoietic response, as well as an unconjugated hyperbilirubinemia (total serum bilirubin: 6.05 mg/dL, unconjugated bilirubin: 5.35 mg/dL) and a lactate dehydrogenase exceeding the laboratory measurable values.

Since extensive evaluation could not find an internal cause, the patient was re-introduced for a paediatric cardiologist consultation. The electrocardiogram showed an atrial sensed, ventricular paced rhythm with occasional ventricular extra beats. Echocardiography revealed moderate tricuspid valve regurgitation due to annuloplasty ring dehiscence from the septal aspect of the valve (Figure 1) as etiologic reason for mechanical haemolysis. This led to surgical indication of re-operation (Figure 2) with multidisciplinary decision for a mechanical valve replacement with a 30 mm SJM-MECJ-502 Mitral 33®. Postoperatively, he was admitted to the ICU, extubated early with only minimal need for catecholamines, and transferred to the paediatric cardiac intermediate care unit two days after the intervention. Haemolysis parameters, including total and unconjugated bilirubin (Figure 3), normalised, and jaundice disappeared within four days after valve replacement. On day five, he was discharged with only mild medication for heart failure and phenprocoumon for permanent anticoagulation.

Discussion

Tricuspid regurgitation is a common complication seen in patients with hypoplastic left heart syndrome, resulting from various congenital and acquired factors like a highly variable subvalvar apparatus, annulus diameter or number of leaflets as well as leaflet dysplasia with preference for the septal one and consecutive movement restriction and higher incidence of cleft formation.[1–3] Throughout the palliation stages, the systemic hypertrophied ventricle evolves a spherical ventricular geometry, which leads to shape-changes of atrioventricular valvular closure as well as to impairment of systolic and diastolic function, which furthermore contributes to tricuspid regurgitation.[4,5] In the past years, atrioventricular valve dysfunction in Fontan patients has become focus of scientific research, indicating higher morbidity and mortality, for example, increased risk of Fontan failure and transplantation in those with

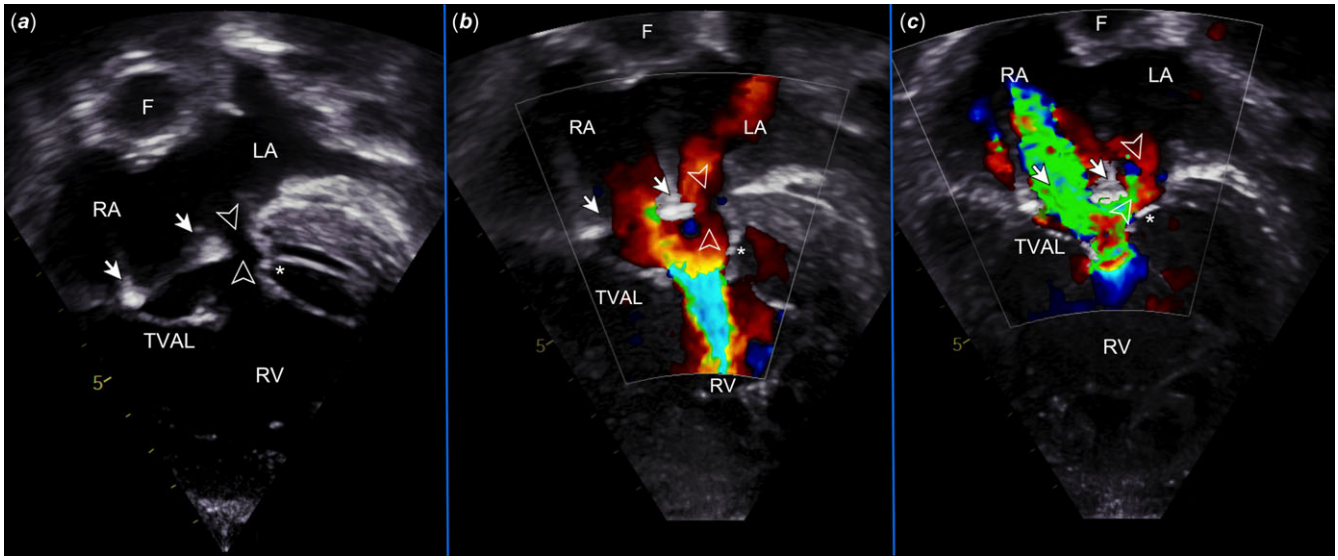

Figure 1. (***a***) Two-dimensional transthoracic echocardiography apical view indicating dehiscence of the tricuspid annuloplasty ring from the septal valve aspect during diastole. (***b***) Colour Doppler image during diastole with blood inflow separated by the septal part of the ring and (***c***) tricuspid regurgitation during systole. F = extracardiac Fontan connection, LA = left atrium, RA = right atrium, RV = right ventricle, TVAL = anterior leaflet of tricuspid valve, *septal leaflet of tricuspid valve, empty arrow heads: area of dehiscence, filled arrows: tricuspid annuloplasty ring.

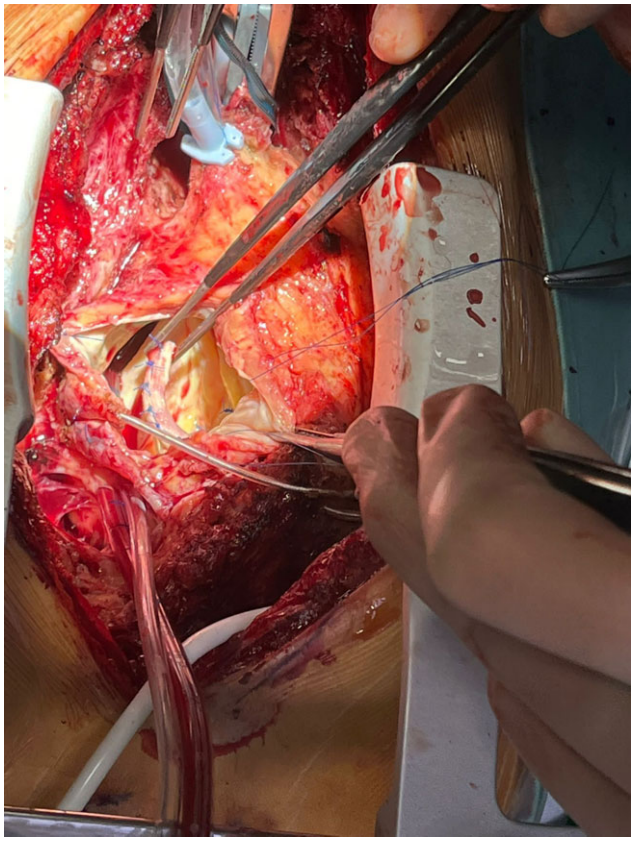

Figure 2. Intraoperative surgeons view showing the loose part of the annuloplasty ring held by the forceps.

moderate or severe regurgitation.[6] Optimal timing and technique for atrioventricular valve intervention still is controversially discussed; it should be mentioned that repair should be aimed for before the development of definitive right ventricular dysfunction, since in these patients, tricuspid valves tend to continue to deteriorate.[7]

Valve-sparing techniques are preferred; the most common ones are (partial) annuloplasty, commisuroplasty, and edge-to-edge repair. Nonetheless, careful preoperative imaging should always precede and contribute to surgical decision-making.[8] Data on tricuspid annuloplasty ring dehiscence in the paediatric cohort are sparse; there are, however, numerous reports on this complication after mitral valve repair in adults.

To the authors' knowledge, there are no reported cases of early tricuspid annuloplasty ring dehiscence presenting with mechanical haemolysis in adolescents with hypoplastic left heart syndrome and Fontan physiology, whereas in adults, haemolysis and hyperbilirubinemia due to failed annuloplasty rings have been reported sporadically.[9]

Our patient recovered fast and was clinically asymptomatic after definitive atrioventricular valve replacement regarding jaundice and bilirubin values.

Despite the lack of guidelines on management and follow-up after atrioventricular valve repair in the reviewed literature, the authors stress that reoccurring postoperative valve regurgitation, with or without rise in haemolysis parameters, should always arouse clinical suspicion of valve repair failure. Three-dimensional transthoracic or conventional transesophageal echocardiography is a promising diagnostic imaging tools with additional benefit in these patients. The annuloplasty ring used in our patient was of rigid nature; experience in adults shows an increased risk of

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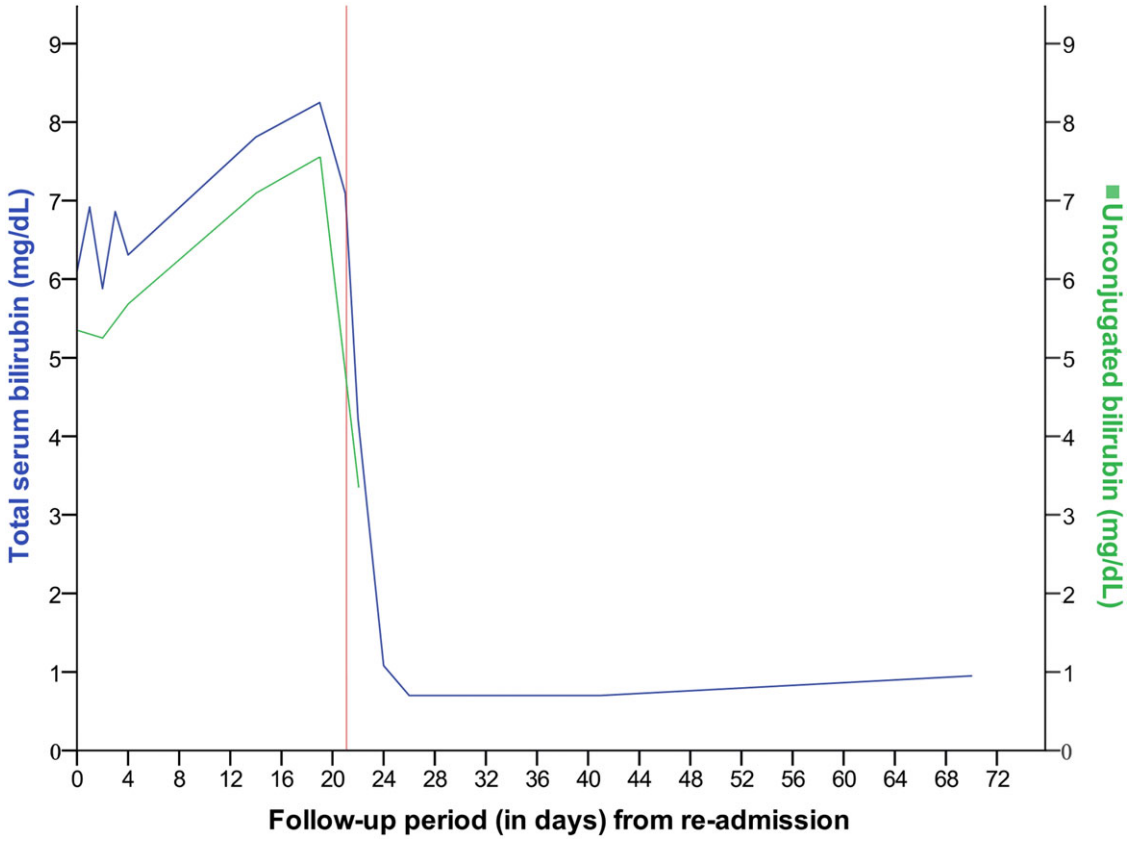

Figure 3. Dynamics of total serum (blue line) and unconjugated (green line) bilirubin (mg/dL) from readmission showing dramatic drop of these parameters immediately after implantation of the mechanical tricuspid valve on day 21 indicated by the red vertical line. Follow-up total serum bilirubin consistently showed normal values until day 70 after readmission; unconjugated bilirubin was too low to be determined.

dehiscence with this kind of ring.[10] Applicability of these data on children with CHD, however, is problematic, as studies are lacking.

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Competing interests. None.

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