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IC001

Interventional Cardiology & Catheterization • Oral Presentation

SUCCESSFUL DEVICE CLOSURE OF A LARGE ATRIAL SEPTAL DEFECT IN A PATIENT WITH EHLER'S DANLOS SYNDROME: A CASE REPORT.

Naseer Kolko, Maryam Usman, Ali Al Akhfash, Abdullah Alqwae, Abdulrahman Al-Mesned

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BACKGROUND: Ehlers-Danlos Syndrome (EDS) is a connective tissue disorder with multisystemic manifestations such as delayed wound healing, skin hyperextensibility, joint hypermobility, recurrent fractures, increased skin fragility and a predisposition to excessive bleeding. While some congenital heart defects have been described in some patients EDS, atrial septal defect (ASD) is relatively rare. Also, there is limited literature on the safety and feasibility of interventional cardiac procedures in this category of patients.

METHODOLOGY: A case of successful device closure of Large ASD in a 7-year-old girl with EDS.

RESULTS: A 7-year-old girl with EDS (confirmed through genetic studies) who has multisystemic manifestations [glaucoma, inguinal hernia, recurrent fractures, hearing impairment, ecchymosis following minor trauma, and poor wound healing after previous surgeries]. She had a screening echocardiography which showed a large ASD. She was referred to center for evaluation and possible transcatheter or surgical closure of the ASD. Physical examination revealed dysmorphic features, including hypertelorism, a high-arched palate, and pectus carinatum, as well as skin hyperextensibility, joint hypermobility, and allodynia. Cardiovascular examination showed a pulse rate of 105 beats/min, blood pressure of 110/65 mmHg, a normal first heart sound, and an accentuated second heart sound. Transthoracic and transesophageal echocardiography confirmed a large secundum ASD measuring 21 mm. Laboratory findings were unremarkable. After a multidisciplinary team discussion, we concluded that transcatheter device closure was the most appropriate intervention and parents were counselled in details for this and they consented. She had a successful transcatheter ASD device closure with no major complications.

Keywords: Atrial septal defect, Device closure, Ehlers-Danlos syndrome, Connective tissue disorder.