Perkutana intervencija na stenoziranom ekstrakardijalnom provodniku u pacijentice s kongenitalnom srčanom greškom korigiranom po Fontanu i dekstrokardijom

Percutaneous intervention on stenotic extracardiac conduit in a patient with Fontan and dextrocardia

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Uvod: Liječenje i praćenje bolesnika s kongenitalnim srčanim greškama u Hrvatskoj bremenito je relativno malim iskustvom i ograničenim dijagnostičkim i terapijskim mogućnostima. Prikazujemo slučaj bolesnice rođene s kompleksnom srčanom greškom koja je korigirana po Fontanu u djetinjstvu, a čiji se tijek bolesti komplicirao opstrukcijom lumena ekstrakardijalnog provodnika postavljenog na srce koje je u dekstrokardiji.

Prikaz slučaja: Bolesnica u dobi od dvadeset i četiri godine hospitalizirana je u Kliničkom bolničkom centru Zagreb zbog srčanog popuštanja koje se manifestiralo otežanim disanjem i oteklinama nogu i trbuha. Radi se o bolesnici koja je rođena s kompleksnom prirođenom srčanom greškom (dekstrokardija, "double inlet" lijevi ventrikul, pulmonalna atrezija i transpozicija velikih krvnih žila). U dobi od mjesec dana učinjena je operacija po Blalock-Taussigu, dvije godine kasnije korekcija po Glennu, a u dobi od osam godina operacija po Fontanu[1]. U redovitoj kontroli, u rujnu 2014. godine, ehokardiografski je prikazan jedan ventrikul (morfološki dominatno lijevi) održane globalne sistoličke funkcije, a ekstrakardijalni provodnik bio je normalnog protoka. Osam mjeseci kasnije bolesnica se prezentirala slikom srčanom popuštanja. Učinjenom višeslojnom CT (MSCT) angiografijom i kavografijom, utvrđena je kalcifikacija i parcijalna tromboza vaskularnog provodnika koji je u središnjem segmentu koljenasto presavinut uz gotovo potpunu opstrukciju protoka. Dva tjedna kasnije, bolesnica je premještena u Herzzentrum u Münchenu. Uslijedila je dilatacija, a potom i postavljanje tri stenta s odličnim finalnim protokom. Kliničko stanje bolesnice postupno se poboljšalo. Kontrolna MSCT angiografija ne nalazi značajnijeg suženja stentiranog ekstrakardijalnog tunela.

Zaključak: Dekstrokardija u bolesnice sa korigiranom srčanom greškom razlogom je postavljanja dugog ekstrakardijalnog provodnika između donje šuplje vene i plućne arterije, a koji je nakon šesnaest godina stenozirao na mjestu koljenastog presavinuća. Učinjena je uspješna perkutana intervencija koja je poštedila bolesnicu potencijalne četvrte kardiotomije.

Introduction: In our country, treatment and follow up of grown-up congenital heart disease patients is limited with relatively low experience and modest diagnostic and therapeutic possibilities. We present a case of a young patient born with complex congenital heart disease corrected with Fontan procedure in childhood, whose medical history was complicated with extracardiac tunnel obstruction.

Case report: Twenty-year-old patient was hospitalized in University Hospital Centre Zagreb for cardiac decompensation manifested with dyspnea, leg edema and ascites. Patient was born with complex congenital heart disease (dextrocardia, "double inlet" left ventricle, pulmonary atresia and great vessels transposition). On her first month, Blalock-Taussig was performed, two years later Glen correction followed and in her eighth year Fontan operation[1] was done. On her regular check-up visit in September 2014, echocardiographic assessment showed her one ventricle (morphologic left) with normal global systolic function and her extracardiac tunnel had normal flow. Eight months later she presented with cardiac decompensation. Multi slice CT (MSCT) angiography and cavography, performed in UHC Zagreb, found calcifications and partial thrombosis of the conduit which was knicked on its mid portion with complete obstruction of the flow. Two weeks later our patient was transferred to Herzzentrum Munich. Dilatation was performed, followed with implantation of three stents and very satisfactory final flow. Clinical status of our patient improved. MSCT angiography showed no residual stenosis on a stented extracardiac conduit.

Conclusion: Dextrocardia, in a patient with complex congenital heart disease, complicated stenting of the extracardiac conduit which was placed between vena cava inferior and pulmonary artery sixteen years earlier and which was knicked on the midportion because it was placed in the heart on the right side. Successful percutaneous intervention was made and three stents implanted. Procedure spared our patient from the fourth cardiotomy.

LITERATURE

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