

Indication for Transplantation in a Patient with Univentricular Heart

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Rezumat

Indicația de transplant la pacientul cu cord univentricular

Corecția chirurgicală a cordului univentricular este o realizare a ultimelor 4-5 decenii. Circulația cavo-pulmonară de tip Fontan este, în ziua de azi, o tehnică chirurgicală care și-a dovedit eficacitatea, dar este în același timp în permanent proces de ameliorare a rezultatelor. În pofida acestui fapt, deteriorarea cu timpul a cordului univentricular este un proces binecunoscut, momentul apariției insuficienței cardiace depinzând de o serie de factori: tehnica chirurgicală, momentul operației, tipul de ventricul unic. Grupul de studiu este reprezentat de 39 de pacienți cu circulație de tip univentricular, operați (37 cu conexiune cavo-pulmonară parțială, 2 cu conexiune cavopulmonară totală) în clinica noastră, în perioada 2006- 2012. Din cadrul acestui grup, 2 pacienți au atins stadiul la care necesită transplant cardiac : unul a fost deja transplantat iar celălalt este pe lista de așteptare. Indicația a fost, în cazul ambilor pacienți, disfuncția ventriculului unic de tip ventricul stâng, în stadiul de conexiune cavo- pulmonară parțială, efectuată la 14 ani de viață. Evoluția la pacientul transplantat este bună. În concluzie, opțiunea de transplant cardiac trebuie să fie accesibilă și avută în vedere la pacientul cu cord univentricular operat, chiar și la vârsta pediatrică, deoarece particularitățile locale determină apariția disfuncțiilor de ventricul chiar mai repede decât ceea ce descrie literatura.

Cuvinte cheie: cord univentricular, transplant, copil

Abstract

Surgical correction of the univentricular heart is an achievement of the last 4-5 decades. The Fontan pathway is nowadays an established but continuously improving surgical technique. Nevertheless, attrition of the univentricular heart is a well-known process, its timing depending on several factors: technique, time of operation, type of ventricle asso. The study group is represented by 39 patients with univentricular circulation, operated (37 partial-PCPC and 2 total - TCPC cavo-pulmonary anastomosis) in our clinic between 2006 - 2012. Out of this group, 2 patients have reached the stage of indication for heart transplantation: 1 has already been transplanted and one is listed for transplantation. The indication was in both a failing ventricle of left-type univentricular heart, in a PCPC status, performed after 14 years of age. Evolution in the transplanted patient is good. In conclusion, the option of heart transplantation should be available and kept in mind for the operated univentricular heart even at pediatric ages, as local peculiarities cause these hearts to fail even earlier than described in the literature.

Key words: univentricular heart, transplantation, child

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Introduction

Therapy of the univentricular heart, with all its anatomical

variants, represents one of the most important achievements in the field of cardiovascular surgery in the last decades. Started in the 60s, the process of understanding the way of functioning of the univentricular heart in order to discover a way of treatment had to first overcome the mental barrier of a heart functioning by other rules than those established for centuries. Gathering of anatomical, functional, haemodynamic and imaging information, followed by the development of the concept of a heart functioning without a subpulmonary ventricle has revolutionized the prognosis of these patients which were, until recently, considered inoperable. But a unitary management of these patients is yet to be reached, one reason being the fact that the „oldest” patients with this type of operation have just now reached their 5th decade of life, while the most adequate way of managing this lesion is changing every year.

The aim of this study was to assess the need for heart transplantation in a group of patients with univentricular heart, which are on the way to have a Fontan pathway established.

Material and Method

Our study group included 39 patients with a univentricular heart physiology, operated in the Institute of Cardiovascular Diseases and Transplantation Tg. Mures between the years 2006-2012. The ages of our patients ranged between 6 months and 14 years; 37 of these patients had a PCPC, while 2 patients went all the way to a TCPC. There were no differences in the operative techniques, as one operator had performed all surgeries. All single ventricles were of left type,

the diagnosis ranging from tricuspid atresia, to double inlet left ventricle. Surgical history before Glenn's had no uniform pattern, as there were patients with Blalock Taussing shunts, banding of the pulmonary artery, aorto-pulmonary fenestration plus banding of the pulmonary artery as well. Assessment before Glenn was performed in a uniform way, including heart ultrasound and diagnostic catheterisation (which followed a common protocol of determining hemodynamics-pulmonary artery pressures and resistances, transpulmonary gradient, Qp/Qs , Rp/Rs , pulmonary artery indices, single ventricle function and establishing anatomy - venous systemic return, veno - venous and arterio - venous collaterals, atrioventricular valve regurgitation, pulmonary artery stenosis). Criteria for performing Glenn were: mean pressure in the pulmonary artery less than 15 mmHg, indexed pulmonary artery resistances less than 3 Wood Units/m², efficient single ventricle, normal pulmonary artery indices, less than moderate arterio-venous and veno-venous collateral, less than moderate regurgitation of the atrioventricular valve. The same assessing protocol and acceptance criteria were used before Fontan operations.

Results

Out of the 39 patients, only 2 had a failing physiology of either the Glenn or the Fontan physiology.

The first patient is a 16 year old female patient with tricuspid atresia and pulmonary hypoperfusion, for which she had previously received two BT shunts, as a small child. At the age of 14, after non-invasive and invasive assessment, she was considered to have an indication for a Glenn and a

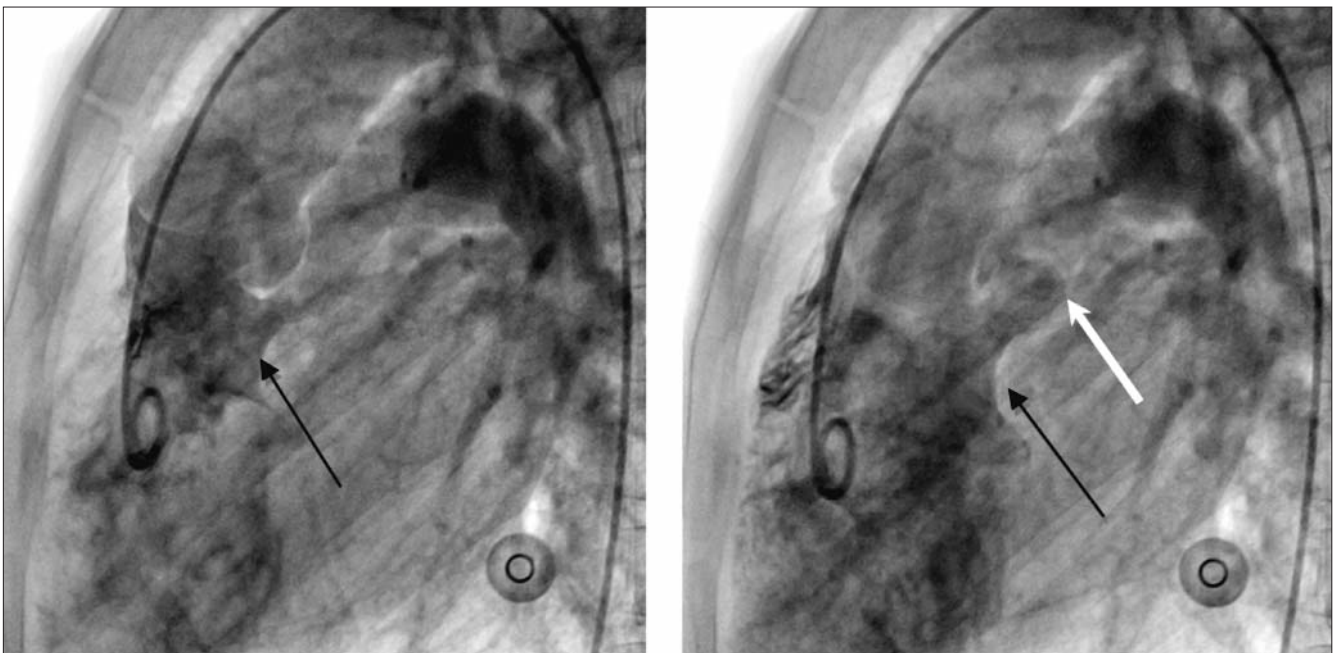


Figure 1. PreGlenn cardiac catheterisation. Angiography, lateral image. Injection in the functional single ventricle (LV), situated anteriorly. Both great arteries emerge from this ventricle: the aorta anteriorly and the pulmonary artery posteriorly. Subvalvular (black arrow) and valvular (white arrow) pulmonary stenosis)

partial cavo-pulmonary anastomosis was performed. Postoperative evolution was good, but after 2 years of hemodynamic stability, her condition worsened, with exercise intolerance, decreasing saturations and worsening function of the single left ventricle on echocardiography. As no surgical treatable lesions were found, she was listed for transplantation and received an isogroup, isoRh heart at the age of 19. No invasive hemodynamic assessment was performed before transplantation. Postoperative evolution on short and medium term is good.

The second patient was diagnosed at birth with tricuspid atresia with transposed great arteries and significant pulmonary stenosis (pulmonary hypoperfusion). He underwent a modified Blalock Taussig shunt operation as an infant and another one at the age of 2 years, due to obstruction of the first shunt. After receiving a 6 mm BT shunt at 2 years of age, his condition was stable for several years, followed by desaturation, exercise intolerance, failure to thrive (weight less than 5th percentile for age). At the age of 12 he was assessed with the described protocol and was considered as having an indication for PCPC. The postoperative evolution was good and the patient was discharged 8 days after the operation. In the next couple of years his condition, initially good, worsened progressively, with marked desaturation exercise intolerance and severe systolic and diastolic dysfunction of the single ventricle on ultrasound, despite supportive medication. For this reasons he was listed for transplantation and is still waiting for a heart transplant. No invasive assessment was performed prior to listing.

Discussions

1. Prevalence of heart transplantation for univentricular hearts. In our group, the prevalence of heart transplantation in patients with univentricular hearts was 5.12%. In the series of d'Udekem (1), 9 out of 305 consecutive Fontan patients have been transplanted (2.95%), representing about half of our series. Age at performing Glenn procedure was significantly lower in their series compared to ours (0.8-2.4 years versus 0.6-14 years). In the whole group of children and young adults (less than 21 years) with transplanted hearts in our center (which is also the experience of the entire country), prevalence of univentricular hearts is 20% (1 out of 5 patients). The latest ISHLT Report for the year 2012 describes a prevalence of CHD among heart transplant recipient between 23-76%, depending on the age category (59% in infants, 56% in children aged 1-10 years and 23% in the 11-17 years group) (2).
2. Risk factors for transplantation in patients with PCPC. Both our failing Glenns were patients having their PCPC performed late (after 12 years), after a longstanding period of significant desaturation. Scheuer et al reports a group of 167 patients having a Glenn operation, out of which only 1 required a heart transplant. It has to be underlined that there is a great discrepancy between our group and the one which

Scheurer et al (3) reported, as their group contains patients in which the Glenn operation was performed before 5 years of age, as opposed to ours, in which age at Glenn is up to 14 years. Both our patients had dysfunctional ventricles as the cause of decompensation. The literature reports mainly patients with normal ventricular function and protein losing enteropathy of plastic bronchitis as candidates for heart transplantation (4,5). It might be again that the specific conditions in which our patients were operated (improper preparation of Glenn with late unloading of the ventricle and prolonged desaturation) could be responsible for this peculiar pattern. From the point of view of their original anatomy, tricuspid atresia with a left-type single ventricle is described in the literature as having a more favourable outcome compared to right - sided single ventricle (as is the hypoplastic left ventricle) (6,7). None of the two patients had a significant atrioventricular valve regurgitation, another risk factor for early failing Glenn's, as mentioned in the literature (3). Altogether, these considerations point to late performed Glenn as the cause for failure and need for heart transplantation.

3. Neither patient showed on echocardiography signs of excessive arterio-venous collaterals or pulmonary hypertension. On the other hand, none of the patients was assessed invasively before transplantation or listing. The intuitive assumption, that a still functioning, even failing univentricular heart circulation can't have prohibitive arterial pulmonary artery hypertension proved to be wrong, according to the literature. Pulmonary hypertension can either exist at the moment of transplantation or develop later, due to several causes, mainly related to the peculiarities of the pulmonary vascular bed in patients with univentricular hearts (8). Besides, a complete hemodynamic evaluation might reveal treatable (medical, interventional or surgical) causes of failing univentricular hearts. But this requires a catheter laboratory equipped to perform interventional therapy in these patients. Under these circumstances, every candidate for heart transplantation should have an invasive assessment performed.

Limitations

The small size of our study group makes generalization of our conclusions impossible. On the other hand, as this group includes the whole experience of a country concerning heart transplantation in children and young adults, valuable conclusions can be drawn.

Conclusions

Heart transplantation in failing Glenn/Fontan patients is relatively high compared to literature, the main reason being single ventricle dysfunction due to old age at Glenn operation. Proper listing for heart transplantation in univentricular hearts should include a complete invasive assessment, with interventional therapy performed if necessary.

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