

Indication for Pediatric Heart Transplantation – Romanian Peculiarities

Sglimbea Anca¹, Toma Roxana¹, Suciu H^{1,2}, Deac R²

¹ Clinic of Cardiovascular Surgery, Tîrgu Mureş, Romania

² University of Medicine and Pharmacy, Tîrgu Mureş, Romania

Background: Heart transplantation is nowadays a well established and successful therapy in both pediatric and adult heart failure. There are several guidelines for adult transplantation, while pediatric transplantation benefits of the indications established in 2007 by the American Heart Association. These are based on western experience and infrastructure. In Romania, this therapy is being performed mainly in the last 5 years, with a small number of patients transplanted.

Methods: We have analyzed the way indication for transplantation has been established in our 5 patients and compared it to the recommendations from the generally accepted guidelines.

Results: There are significant differences regarding age at transplantation, underlying heart disease, as well as stage of heart failure and medical therapy before transplantation. Our patients are older, transplanted mainly for dilated cardiomyopathy, mainly in NYHA functional class IV, but not hospitalized or on intensive care.

Conclusion: There is an imperative need for establishing the number of necessary donor hearts at a national level, as well as starting a vigorous campaign to increase organ donation in Romania.

Keywords: heart transplantation, child, guidelines

Received: 5 May 2012

Introduction

Heart transplantation is considered since the '80th a successful but ultimate therapy for heart failure in both children and adults. Still, it is a palliation rather than a curative therapy. As such, in pediatric patients, this palliation is expected to offer the child a lifespan as close as possible to that of a healthy person and with a quality of life similar to what we call normal. That calls for a thorough assessment of the transplant candidate and furtheron, the transplanted patient, in order to create the best conditions for a long term good survival. Indications and management of heart transplantation in children are tailored nowadays to achieve this goal. However, as it often happens, guidelines for the management of these patients are established in countries/medical systems with a much higher financial potential [1,2,3]. That rises the question if those guidelines are completely superposable with specific conditions in other areas of the world. The American Heart Association has released in 2007 the latest and solely guidelines for heart transplantation in pediatric heart disease [1]. In Romania, pediatric heart transplantation is performed, in a rather unsustained rhythm, since 2000. The aim of this study was to analyze our group of pediatric heart transplant patients and compare it with data reported from registries or associations caring for the same type of population.

Material and method

We have retrospectively analyzed our group of pediatric heart transplant recipients. First heart transplantation in Romania was performed in 2000 and last in 2011. Dur-

ing this period there were 4 pediatric (0–18 years) patients receiving a heart, all female. There was one more female patient, 19 years of age, at the limit of our age group. As our center is the only one performing this type of intervention in children in Romania, these group si representing all pediatric heart transplant patients operated in Romania. There are some Romanian children operated for a heart transplantation in other countries in Europe, but the analysis of those patients is beyond the scope of this study. Data collected for these patients were: age at transplantation, period spent on the waiting list, underlying heart disease, indication for listing, heart failure class at transplantation, medication, outcome. As the number of patients in our group is small, data were presented in a descriptive manner.

Results

The results are presented in Table I.

Age at transplantation: the majority of our patients were teenagers.

Period on waiting list: period on waiting list is around 12 month for the listed patients. These are mainly the older children. Infants and small children are not even listed, as consensus about their management (indication, assessment for listing, availability of a donor heart) is poor in Romania.

Most (4 out of 5) of the transplanted children have, as an underlying heart disease causing heart failure, dilated cardiomyopathy. There was only one, almost adult patient, with a congenital heart disease transplanted, a single-ventricle-type heart abnormality after previously performing partial cavo-pulmonary anastomosis at a rather old age for these operation.

Table I. Characteristics of transplanted patients

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age at transplantation	10 years	17 years	15 years	19 years	3 ½ years
Period on waiting list	Not listed	10 month	12 month	10 month	Not listed
Heart disease	DCMP	DCMP	DCMP	TA with PCPC	DCMP
NYHA class of HF	IV	III	IV	IV	IV
AHA stages of HF	D	C	D	C	C
Hospitalized/ outpatient	Hospitalized				
transferred	Outpatient	Hospitalized	Outpatient	Outpatient	
Medication	erratic	Not maximal	Maximal	Not maximal	Not maximal
Outcome	Death 2 years post-op	Alive	Alive	Alive	Alive

DCMP – dilated cardiomyopathy; HF – heart failure; PCPC – partial cavo-pulmonary connection, TA – tricuspid atresia, AHA/ACC – American Heart Association

Functional class heart failure was IV NYHA in 4 out of 5 patients. On the other hand, based on the AHA/ACC classification of heart failure, 3 out of our 5 patients were in stage C. This correlates with the inpatient/outpatient status, as 2 patients were hospitalized, while 3 were at home before transplantation.

Therapy was maximal only in the one hospitalized patient who was waiting for transplantation in our ICU. Outcome was good in the last 4 patients, who were operated in the last 5 years. The first patient died 18 month after transplantation, due to an acute illness, in another hospital.

Discussions

Number of patients

Compared to western countries, the number of transplanted children in Romania is extremely small. What would be worth to know is the necessity of donor hearts, respectively, the true number of pediatric patients on the heart transplantation waiting list. As some categories (infants, small children) are not even listed, due to anterior experience of lack of hearts and lack of this type of procedure in Romania, the true necessity is not even known. In the US, the number of children on the heart transplant waiting list was around 800 in 2009. If extrapolating these data to our country, we would expect to have, if data were comparable, at least 50 children waiting for a heart transplantation each year. But, while in the US about 360 children are transplanted every year (about half of the patients needing a heart transplant) (about 500 children removed from the transplant list each year: about 360 transplants, 80 death while on transplant list, about 50 improved and about 25 too sick to transplant) [5], in Romania the rate of transplantation is less than 1 child per year. That results in a constant residual number of untransplanted alive children of about 1/3 of the necessity per year in the US, compared to about 2/3 in Romania [5].

Age at transplantation

The majority of our transplanted children are teenagers, while in the US, in 2009, the highest number of trans-

plants was performed among infants (less than 1 year of age), followed by the age group 12–18 years and 1–5 years [5]. This is mainly due to the lack of awareness of heart transplantation possibilities nowadays, as well as lack of education of the general population toward donating organs. An important role is also played by the difficulties, in Romania, of declaring patients brain-dead for the category of 0–5 years, as criterias and technical possibilities are limited.

Disease

Conditions considered for pediatric heart transplantation worldwide include: all types of cardiomyopathies, rarely-anatomically uncorrectable congenital heart diseases (especially HLHS), congenital heart disease at high risk for repair (eg, severe Shone complex), refractory heart failure after previous cardiac surgery, significant cardiac allograft vasculopathy or chronic graft dysfunction of a previous heart transplant, unresectable symptomatic cardiac neoplasms [4]. Unfortunately our medical thinking in Romania is only directed toward cardiomyopathies as an indication for heart transplantation, this being the reason for the disease distribution in our study group. One is rarely thinking of heart transplantation as a therapeutical method for a severe Shone complex, for example.

Listing for transplantation

Criterias for listing for transplantation in children in Romania are tending to be taken over from the AHA/ACC guidelines, 2007 [1]. These list as class I criterias a variaties of conditions (stage D HF, but also stage C HF with functional NYHA class IV, growth failure, near- sudden death or untractable arrhythmias aso), far away from the urgency described by the UNOS waiting list (status IA- on ventilator/ECMO/balloon pump/PGE/continous inotropes, life- threatening arrhythmias; status IB- less than 6 month of age, on low- dose inotropes, failure to thrive, aso) [5]. Very few of our patients are in a UNOS IA or IB status, as, if they get into that condition, they die before transplantation. Reasons for this is lack of availability of ECMO or any other ventricular assist device and even, in many centers, of mechanical ventilation or the possibility of admin-

istration of continuous inotropic drugs. This would suggest a rethinking/underlining of stage C heart failure as major transplantation indications in Romania, maybe with a risk stratification based on several demonstrated risk factors [4].

Maximal therapy is rarely achieved in our patients. This is mainly due to the fact that the term of maximal therapy is rarely completely understood, but also because of the general, even medical belief that medical therapy is of not much use in heart failure. General, both medical and parental education is needed in order to achieve the goal of reaching maximal therapy in a child with heart failure.

Conclusion

Pediatric heart transplantation in Romania is an ongoing process which needs to be oriented toward the particular needs of our pediatric population, which seem to be somehow different from those of other western countries from which guidelines have emerged. This calls for a local redefining of assessment, indication and management of heart

failure in children, in order to offer the best solution for this important part of our little heart patients.

Acknowledgement

This paper is partly supported by the Sectorial Operational Programme Human Resources Development (SOP HRD), financed from the European Social Fund and by the Romanian Government under the contract number POSDRU/89/1.5/S/60782.

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