EPIDEMIOLOGIC AND THERAPEUTIC FEATURES OF CRITICAL CONGENITAL HEART DISEASES- A SINGLE CENTER EXPERIENCE

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Abstract: *The progress achieved in the last decades in pediatric cardiology results in a significant decrease in mortality due to congenital heart disease (CHD). In Romania there are no specific data concerning this subject. The authors present a three year experience of a single pediatric cardiovascular center regarding epidemiology, diagnosis and therapy of patients with critical CHD, in a prospective study. The results show a significant percentage of critical CHD (aprox. 20%), with a mean age at diagnosis of 23.6 days. The number of patients undergoing surgical treatment and the results of therapy highlight, the need of early diagnosis and timely refferal for surgical treatment. Conclusion: Critical CHD represent an important part of the pathology of a pediatric cardiovascular center. Nowadays, late age of referral worsens the prognostic of this patients. The necessity of establishing a coherent strategy emerges, in order to offer these children a chance.*

Cuvinte cheie: *malformaţii cardiace congenitale critice, epidemiologie*

Rezumat: *Evoluţia cardiologiei pediatrice în ultimele decenii se concretizează la nivel mondial în scăderea semnificativă a mortalităţii prin malformaţii cardiace congenitale (MCC). România nu dispune de date în acest sens. Autorii prezintă experienţa unui singur centru cardiovascular pediatric în ceea ce priveşte caracteristicile epidemiologice, de diagnostic şi terapeutice ale pacienţilor cu MCC critică, sub forma unui studiu prospectiv, desfăşurat pe o periodă de trei ani în instituţia noastră. Rezultatele arată o pondere semnificativă a MCC critice (cca 20%), cu o vârstă medie la diagnosticare de 23,6 zile, în condiţiile unei patologii complexe. Numărul de pacienţi operaţi şi rezultatele terapiei subliniază necesitatea diagnosticării şi referirii în timp pentru operaţie a acestor pacienţi. Concluzii: Malformaţiile cardiace congenitale critice reprezintă o pondere importantă în patologia unui centru cardiovascular pediatric. În prezent, referirea tardivă umbreşte prognosticul acestor pacienţi. Este necesară stabilirea unei strategii coerente pentru a acorda şansa cuvenită acestor copii.*

INTRODUCTION

The last three decades encountered substantial achievements in the management of congenital heart diseases (CHD), with a reduction of 39% in mortality due to this pathology in western countries and North America.

One of the most important steps responsable for this significant decrease of mortality in CHD is represented by lowering of the age for corrective surgery to the neonatal period and the fundamentation of the term of critical CHD (a CHD necessitating for survival surgical or interventional therapy in the first month of life) (1).

Despite these facts, in the same countries, CHD remain responsible for about 30% of death caused by congenital defects and of 5.7% of infantile mortality, while 57% of these death are encountered during the first month of life (1,2).

In Romania no such data are available.

To complete this lack of information, we are presenting a single center experience, that of the pediatric cardiovascular center (including the 3rd Department of Cardiology and the 2nd Department of Pediatric and Adult Cardiovascular Surgery), part of the Emergency Institute of Cardiovascular Diseases and Transplantation (IUBCvT)Tg. Mures, regarding epidemiology and management of the patient with critical CHD.

MATERIAL AND METHODS

This is a prospective, descriptive study, running between November 2007 and October 2010 in the 3rd Department of Cardiology, a hospital with a substantial number of patients, all with a heart condition, with a mean of 1400 admittances/year. In this three year interval a group of 1249 neonates and small infants, coming from all counties in Romania have been assessed. The evaluation was performed in an outpatient manner, these children being admitted to the Ist Department of Neonatology or the Intensive Care Department of the IUBCvT. The protocol of assessment included clinical exam (3), measurement of non invasive blood pressure (right arm) with a monitor attached cuff, measurement of peripheral oxygen saturation by means of a pulse oxymeter attached to the same monitor (4,5,6,7), 12 leads ECG and echocardiography, performed with a Philips iE33 machine. The echocardiographic exam followed the generally used protocol, with the description of situs, position of the heart, segmental analysis of cardiac structures, assessment of pressures in different parts of the heart and, if needed, calculation of shunts.

The parameters followed were: age at the time of diagnosis, the county of birth, diagnosis, critical nature of the CHD, treatment (operated or not), results of the treatment (survival), if prenatal diagnosis was available.

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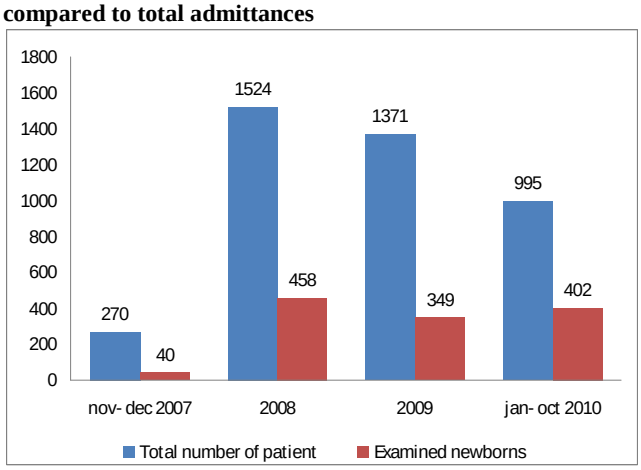
RESULTS

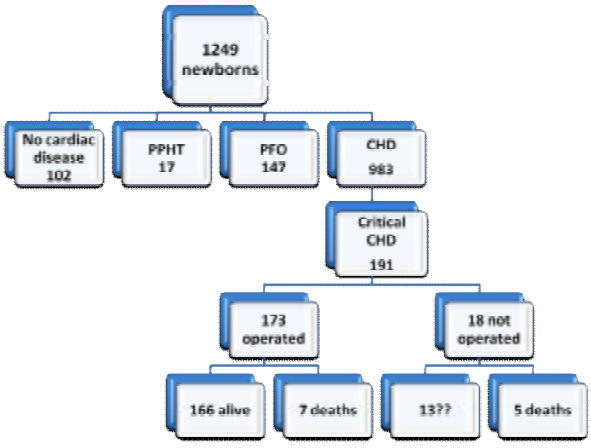
In the period between November 2007 and October 2010 a number of 1249 neonates and small infants was assessed in our department, with an average of 415,6 cases/year. The distribution per years of the assessed cases is represented in Fig. 1, compared to the total number of cases admitted in the same period in our department. Distribution per regions shows 75 cases coming form the southern part of the country (Muntenia, Oltenia, Dobrogea), 75 patients form central and western Romania (Transilvania, Banat) and 41 patients form the eastern part (Moldova).

Figure no. 1. Distribution per years of the neonates compared to total admittances



From the total of 1249 examined children (Fig. 2), 102 had no heart condition, 17 had persistent pulmonary hypertension of the newborn and 147 a patent foramen ovale. The rest of 983 patients were diagnosed with CHD.

Figure no. 2. Composition and evolution of the study group



191 of the studied patients (19.43%) were diagnosed as having a critical CHD. Distribution by lesions is presented in Table II (divided upon type of surgical correction as the number of ventricles and LV outflow tract obstruction are prognostic elements for these patients.

Age at diagnosis was 1 to 84 days (mean 23,59 days). 173 patients underwent surgery, of which 166 are alive. 7 patients have died (5 transpositions of the great arteries, 1 coarctation of the aorta and 1 patient with a single ventricle physiology).

Out of the 18 patients not operated, 5 died (2 with a hypoplastic left ventricle, 3 with transposition of the great arteries), while the rest of 13 are lost for follow-up.

During this period, a number of 41 pregnant women have been reffered for fetal echocardiography, with a suspected CHD in the fetus. In 31 fetuses, CHD has been confirmed, out of which 15 were delivered in our center and available for further follow-up.

Table no. 1. Distribution of the cases according to the type of CHD

Type of CHD		Number of the cases	Total
CHD with left-to-right shunt		714	1249
Complex CHD	TGA	82	
	AoCo	25	
	AoCo + CAVSD	4	
	TF	25	
	CAVSD	27	
	AoS	15	
	PAt	15	
	PS	10	
	TAt	7	
	Ebstein anomaly	7	
	TAPVR	7	
	TAC	6	
	Shone syndrome	2	
	Univentricular heart	30	
PPHT		17	
HCM		7	
Innocent heart murmur/no modification		102	
PFO		147	

Legend: AoCo= coarctation of the aorta; AoS= aortic stenosis; CAVSD= complete atrioventricular septal defect; PAt= pulmonary atresia; PS=pulmonary stenosis; TAPVR= total anomalous pulmonary venous return; TAC= truncus arteriosus communis; TF= tetralogy of Fallot; TGA= transposition of the great arteries; TrA= tricuspid atresia; PPHT= persistent pulmonary hypertension; PDA=persistent ductus arteriosus; ASD=atrial septal defect.

Table no. 2. Distribution of the critical CHD according to the type of lesions

Biventricular heart with LV obstruction		Total	%	Univentricular heart with LV obstruction		Total	%
IAA	1	35	18,32%	HLHS	8	11	5,75%
AoC	25						
AoS	8			DILV	3		
CAVSD+AoS	1						
Biventricular heart without LV obstruction		**Total**	**%**	**Univentricular heart without LV obstruction**		**Total**	**%**
PAt	2	122	63,87%	PAt	14	23	12,04%
AVSD	4			TAt	5		
AVSD + PAt	2			TS	3		
DORV	13						
PS	6						
TAPVR	4			TGA anatomically corrected	1		
Ebstein anom.	1						
TAC	1						
TF + PAt	5						
TF +P agenesia	1						
TGA	83						
TOTAL 191 cases							

Legend: LV= left ventricle; IAA= intrerrupted aortic arch; AoCo= coarctation of the aorta; AoS= aortic stenosis; AVSD= complete atrioventricular septal defect; HLHS=hypoplastic left heart syndrome; DILV= double inlet left ventricle; PAt= pulmonary atresia; DORV=double outlet right ventricle; PS=pulmonary stenosis; TAPVR= total anomalous pulmonary venous return; TAC= truncus arteriosus communis; TF= tetralogy of Fallot; TGA= transposition of the great arteries; TrA= tricuspid atresia; TrS= tricuspid stenosis.

DISCUSSIONS

Critical CHD represent a major health problem in the neonatal pathology, as it represent over 20% of all CHD diagnosed and treated in this age group in a tertiary center (an average of 64 patients per year). Under current organizing conditions, this number is probably underestimated, as we receive daily a significant number of requests for admitting patients form all over the country, which we can't admit and treat entirely.

As we are speaking about an emergency pathology, and as early establishment of the diagnosis is vital, a mean age at diagnosis of about a month (23,59 days), at which our patients have been diagnosed is completely unacceptable.

In Romania, fetal echocardiography is at it's early beginnings, thus the percentage of patients with a fetal diagnosis being very small (1.52% of the CHD in our study group). There is a huge possibility of diminishing this problem, by establishing an assessment protocol of the pregnant woman with description of risk groups necessitating a mandatory fetal echocardiography performed by a pediatric cardiologist, training the obstetricians in the knowledge of CHD pathology, physiology and echocardiographic diagnosis, aso.

The need for a screening program for CHD in the neonatal period becomes obvious. This program should include clinical exam and pulse oximetry (8), allowing timely diagnosis and referral to a diagnostic center, followed, if needed, by the transfer to a pediatric cardiovascular center. Reasons for such a program are found both in medical literature (as the incidence of critical CHD is established at 1 per 15000- 26000 live birth and most of the diseases for which a screening program is running qualify in these limits) (1,9,10) and in our study (outlining the delayed diagnosis of this children). Delayed diagnosis is usually associated with significant hemodynamic and metabolic compromise, causing neurological and infectious complications and an increase in mortality, diminishing the prognosis of these children.

If diagnosed correctly, results of surgery in these patients are extremely favorable, with a mortality approaching, in our center, the data cited by the literature for important hospitals, despite the presence of a significant number of patients with risk factors (univentricular heart, LV outflow tract obstruction).

Briefly, directions to follow are: increasing fetal diagnosis, screening for CHD in neonates in nurseries, development of diagnostic centers for establishment of a correct diagnosis and recommended therapy, proper financial support for pediatric cardiovascular centers, so that these can cover the necessary treatment for this category of patients.

CONCLUSIONS

Critical CHD represent an important part of the pathology of a pediatric cardiovascular center. Nowadays, late age of referral worsens the prognostic of this patients. The necessity of establishing a coherent strategy emerges, in order to offer these children a chance.

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