CLINICAL STUDY

Neonates with left-sided obstructive heart disease: clinical manifestation and management at primary care hospitals

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Abstract

Background: **In neonates, left-sided obstructive heart defects are critical diseases requiring early recognition, initial stabilization, and transfer to Cardiac Center.**
Objectives: **To assess management of these neonates in primary care hospitals.**
Methods: **Retrospective analysis of medical records of neonates admitted to Cardiac Intensive Care Unit in whom diagnosis of left-sided obstructive heart defect was established.**
Results: **During 8-year period records of 117 patients were evaluated. Of those, 83 had hypoplastic left heart syndrome, 13 aortic stenosis, 12 coarctation of aorta, and 9 interruption of aortic arch. In 13 (11 %) patients diagnosis of heart defect was established prenatally, other neonates presented postnatally with heart failure (28.2 %), shock (65.4 %) or severe cyanosis (4.8 %). Continuous infusion of PGE1 and dopamine was administered in 84.6 % and 30.8 % patients, respectively. Thirty-two (30.8 %) patients were mechanically ventilated and 36 (34.6 %) received sodium bicarbonate. In referring hospitals the heart defect was most often diagnosed using echocardiography and diagnosis was established accurately in 55 % patients.**
Conclusion: **The study showed that in primary care hospitals neonates with known or suspected left-sided obstructive heart disease need more aggressive resuscitative measures than those used in this patient series. Prenatal diagnosis rate of these heart defects is very low *(Tab. 3, Fig. 1, Ref. 8)*. Full Text (Free, PDF) *www.bmj.sk*.**
Key words: congenital heart defect, left-sided obstructive heart disease, neonates.

Left-sided obstructive heart diseases represent a spectrum of abnormalities of the left vetricular inflow (mitral valve), cavity (ventricle), outflow (subaortic area, aortic valve), and aortic arch. In critically ill newborns, these diseases relate to four major diagnostic categories: hypoplastic left heart syndrome (HLHS), aortic stenosis (AS), interrupted aortic arch (IAA), and coarctation of aorta (COA) (with or without associated ventricular septal defect) (1). The lesions present with congestive heart failure, shock, or less commonly with cyanosis, and require early active medical management. To achieve favourable outcome multidisciplinary approach with cooperation of neonatologists, pediatric cardiologists, intensivists and cardiosurgeons is required. In Slovakia, all critically ill neonates with suspected congenital heart disease (CHD) are transferred to Pediatric Cardiac Center in Bratislava where specialized care for the whole spectrum of congenital heart diseases is provided.

The aim of this study was to assess the clinical presentation and management of neonates with left-sided structural heart diseases at neonatal units of primary care hospitals prior to their admission to the specialized center for heart diseases.

Patients and methods

Medical reports of neonates with left-sided obstructive lesions admitted to Pediatric Cardiac Intensive Care Unit (PCICU) between January 1997 and December 2004 were evaluated retrospectively. Cardiac anatomic inclusion criteria included neonates with: a) aortic stenosis, b) hypoplastic left heart syndrome, c) coarctation of aorta (with/without ventricular septal defect) and d) interrupted aortic arch. Neonates with other, more complicated forms of congenital heart diseases were excluded from

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Tab. 1. Characteristics of patients.

	Hypoplastic left heart syndrome (n=83)	Aortic stenosis (n=13)	Coarctation of aorta (n=12)	Interrupted aortic arch (n=9)
Male/Female	50/33	10/3	7/5	6/3
Weight (g)	3230	3500	3215	3350
median (range)	(1700–4200)	(1880–4110)	(2870–4160)	(1800–4000)
Associated noncardiac anomalies	10	2	2	5
Genetic anomalies				
Dysmorphism	5	0	0	0
Named genetic syndrome	2	2	0	5

Tab. 2. Clinical presentation and initial management of newborns with postnatally diagnosed congenital heart defect.

	Hypoplastic left heart syndrome (n=74)	Aortic stenosis (n=9)	Coarctation of aorta (n=12)	Interrupted aortic arch (n=9)
Age at presentation (days)	2 (1–14)	1 (1–4)	13 (1–22)	3 (1–4)
Symptoms:				
Heart failure	23	2	2	2
Shock	47	7	8	6
Profound cyanosis[a]	2	0	2	1
Renal dysfunction[b]	39	9	7	7
Metabolic acidosis[c]	49	6	8	6
Highest recorded base deficit[d] (mmol/l)	8.0 (4.4–29)	7.0 (5.1–19)	18.8 (5.6–26)	9.2 (5.4–10)
Initial management:				
PGE1	63	8	8	9
Catecholamines	20	4	5	3
Mechanical ventilation	22	4	5	1
Sodium bicarbonate	24	3	8	1

[a] – arterial saturation O_2<60 %, [b] – serum creatinine >80 μmol/l, [c] – defined as a base deficit >4 mEq/L, [d] – in patients with metabolic acidosis

our analysis. Demographic data, clinical presentation of heart defect, cardiac diagnostic evaluation in the referring hospital, prenatal establishment of diagnosis, associated anomalies, and initial resuscitative management before admission to PCICU were assessed.

Data are presented median values (range).

Results

During the 8-year period (1997–2004), 150 neonates with the above mentioned diagnosis were admitted to PCICU. 33 patients were eliminated from our analysis due to data incompleteness. Medical records of 117 patients were evaluated; 83 patients had HLHS, 13 patients had AS, 12 patients had COA, and 9 patients had IAA. The number of males was higher than that of females in the whole group of patients, as well as in all diagnostic subgroups. Birth weight in this series of patients was 3280g (1700–4200 g). Anomalies affecting other than the cardiovascular system occurred in 19 (17.1 %) patients; 9 (7.7 %) patients had named genetic anomalies, and 5 (4.3 %) patients demonstrated dysmorphism. The highest incidence of genetic anomalies was found in patients with IAA; all represented DiGeorge syndrome. Demographic data and incidence of associated anomalies are listed in Table 1.

In 13 (11 %) of 117 newborns the diagnosis of congenital heart defect was established prenatally; these babies were delivered at Maternal-Fetal Medicine Center at University Hospital Bratislava, and transferred to PCICU immediately after birth. In other 104 (88.9 %) newborns heart lesion manifested 2 days (0–22 days) after birth in 29 (28.2 %) of these patients CHD manifested with heart failure; in 68 (65.4 %) with shock, and in 5 (4.8 %) with profound cyanosis. Renal dysfunction (defined as serum creatinine 80 mol/l) occurred in 62 (59.6 %) patients. Sixty-nine (66.3 %) patients demonstrated metabolic acidosis defined as base deficit 4; the highest recorded base deficit was 8.0 (4.0–29). Eighty-eight (84.6 %) patients received continuous infusion of PGE1, 32 (30.8 %) continuous infusion of catecholamines, and 36 (34.6 %) patients received boluses of sodium bi-

Tab. 3. Tests used in diagnostic evaluation of congenital heart disease prior to transfer to Pediatric Cardiac Center.

	Hypoplastic left heart syndrome (n=83)	Aortic stenosis (n=13)	Coarctation of aorta (n=12)	Interrupted aortic arch (n=9)
Prenatal echocardiography	9	4	0	0
Postnatal evaluation:				
Chest radiograph	52	9	9	9
Electrocardiogram	27	9	4	4
Echocardiography	54	12	8	8

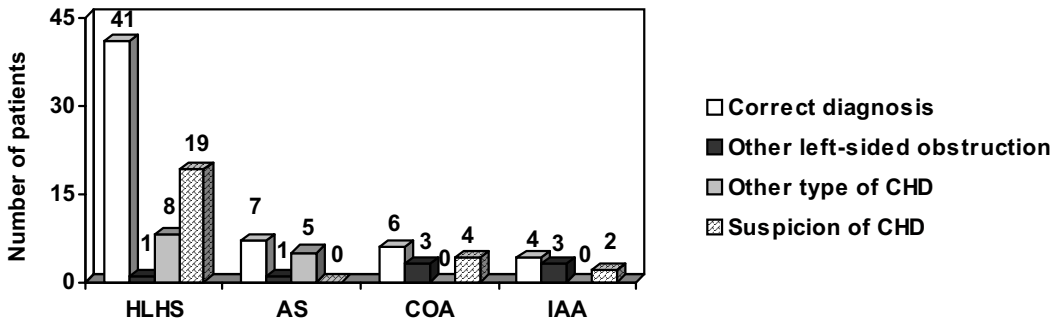

Fig. 1. Accuracy of diagnoses established prior to admission to Pediatric Cardiac Center. CHD – congenital heart defect, HLHS – hypoplastic left heart syndrome, AS – aortic stenosis, COA – coarctation of the aorta, IAA – interrupted aortic arch.

carbonate. Thirty-two (30.8 %) patients were mechanically ventilated. The clinical manifestation of individual CHD and therapeutic measures used for the initial stabilization of newborns are shown in Table 2.

Sixty-nine patients (66.3 %) with postnatal diagnosis of CHD were transferred from their primary care hospital to PCICU directly, and 35 patients (33.7 %) indirectly (through tertiary care hospital).

Tests used for cardiac diagnostic evaluation before the admission to Cardiac Center and the accuracy of the established diagnosis are shown in Table 3 and Figure 1, respectively.

Discussion

Critical left-sided structural heart diseases manifest as a life-threatening condition at neonatal age. Generally, these babies appear normal at birth; problems occur when the ductus arteriosus begins to close and they result in systemic hypoperfusion, congestive heart failure, shock, risk of renal insufficiency, hepatic dysfunction and necrotizing enterocolitis. The closure of ductus arteriosus leads to hypoperfusion of the lower half of the body in COA and IAA, and to hypoperfusion of the entire systemic circulation in AS and HLHS (1). Patients may be also deeply cyanotic because of small interatrial communication and inadequate pulmonary blood flow (2). In this series, 65.4 % of the patients presented with shock, 28.2 % with heart failure and 4.8 % with profound cyanosis. The clinical presentation is related to the rapidity of ductal closure and also to the severity of left-sided obstruction. We observed a very early manifestation of AS and IAA; in all patients, their disease manifested within the first four days of life. Clinical presentation of HLHS and COA was within a broader range (14 and 22 days, respectively). The median age at presentation of HLHS was much lower than that of COA (2 vs 13 days).

Beside physical examination, initial evaluations of neonates with suspected congenital heart defects should include radiographic, electrocardiographic and echocardiographic evaluations. The most common diagnostic tests used in this cohort were chest radiography and echocardiography; electrocardiography was the least available test at primary care hospitals. Precise diagnoses were established in 55 % of cases.

When the diagnosis of left heart obstructive lesion is confirmed or suspected, the newborn should be stabilized and transferred directly to the Pediatric Cardiac Center. Sixty-six percent of the patients were transferred from primary care hospitals to our center directly; however, 34 % patients were primarily transferred to a secondary or another tertiary care hospital. Such transfer was performed either because CHD was not suspected or because there was an effort to verify the diagnosis of CHD.

The initial stabilization of neonates with left heart obstructive lesion includes resuscitative measures and the effort to main-

tain the patency of the ductus arteriosus with intravenous infusion of PGE1. Although crucial to all patients with ductal-dependant circulation, PGE1 was administered only to 84.6 % of the patients. Endotracheal intubation and mechanical ventilation are indicated for poor respiratory effort, severe hypoxemia, low cardiac output, hemodynamic instability, and apnoe (side effect of PGE1). The transport distance, skill of transport team in emergent intubation, and birth weight should be also considered. Even if shock and severe cyanosis were to be considered to be the indication criteria, the number of mechanically ventilated patients is low.

In neonates with left heart obstructive lesion, volume resuscitation, inotropic support with dopamine, and correction of metabolic acidosis are usually required to improve cardiac output and tissue perfusion (3). Again, considering a number of patients presenting with shock, the administration of catecholamines does not seem appropriate.

A better way to manage a newborn with critical heart defect is to diagnose the defect prenatally. Fetal assessment of heart defect has several advantages including delivery near Pediatric Cardiac Center, prompt initiation of prostaglandin therapy, urgent management of physiologic abnormalities (e.g. restrictive foramen ovale in HLHS), avoidance of the potential complications associated with transport and close proximity to the mother (4, 5). The family may be educated before the delivery, so they can make their decision about the future management of their fetus. In infants with fetal diagnosis, better preoperative condition can be seen, as evidenced by less metabolic acidosis and better renal function (6, 7). Delayed diagnoses of CHD worsen the preoperative condition and outcome of surgery in neonates (8). Fetal echocardiography allows the diagnosis of HLHS, severe AS, large ventricular septal defect; however, prenatal detection rate of CHD was only 11.1 % in this series.

Conclusion

As the most common clinical presentation of left-sided structural heart diseases in neonates is shock, beside the administration of PGE, the low cardiac output in these patients should be treated. This study showed that neonates at primary care hospitals need more aggressive resuscitative measures than those used in this patient series. Great effort should be also exerted to achieve a higher prenatal diagnosis rate.

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