II. NEONATOLOGY

THE PREVALENCE OF THE HEART CONGENITAL MALFORMATIONS TO THE PREMATURE NEW BORN

Daniela Iacob[1], RE Iacob[1], Marioara Boia[1], Aniko Manea[1], Mirabela Dima[1]
[1]University of Medicine and Pharmacy „Victor Babes" Timisoara, Romania

Abstract

Congenital heart diseases occur in approximately 1% of live-born infants and represent an important problem in pediatry. The objectives of this study are to establish the incidence of heart congenital malformations when compared to the other congenital malformations and their frequency according to some factors: risk, social background, sex and prematurity.

Key words: heart congenital malformations**,** prematurity**.**

Introduction

Heart congenital malformations represent an important problem in pediatry because of the growth incidence and of the medical and social implications.

The diagnostic dilemma of the newborn with congenital heart disease must be resolved quickly since therapy may prove lifesaving for some of these infants. Congenital heart disease occurs in approximately 1% of live-born infants. Nearly half of all cases of congenital heart disease are diagnosed during the first week of life. The most frequently occurring anomalies seen during this first week are patent ductus arteriosus, transposition of the great arteries, hypoplastic left heart syndrome, tetralogy of Fallot, and pulmonary atresia.

Symptoms and signs in newborns with heart disease permit grouping according to levels of arterial oxygen saturation (cyanotic heart disease and acyanotic heart disease). Further classification (based on other physical findings and laboratory tests) facilitates delineation of the exact cardiac lesion present.

Cyanotic heart disease

Infants with cyanotic heart disease are usually unable to achieve a PaO_2 above 100 mm Hg after breathing 100% inspired oxygen for 10-20 minutes.

Because of intracardiac right-to-left shunting, the newborn with cyanotic congenital heart disease (in contrast to the infant with pulmonary disease) is unable to raise the arterial saturation, even in the presence of increased ambient oxygen.

Care must be taken in evaluating cyanosis by skin color, since polycythemia, jaundice, racial pigmentation, or anemia may make clinical recognition of cyanosis difficult.

The infant with cyanotic congenital heart disease often does not have a distinctive murmur. In fact, the most serious of these anomalies may not be associated with a murmur at all.

Cyanotic infants may be further classified on the basis of pulmonary circulation on chest x-ray and electrocardiographic findings.

The most frequent cyanotic heart disease abnormalities are:

1. Transposition of the great arteries

Transposition of the great arteries is the most common cardiac cause of cyanosis in the first year of life, with a male/female ratio of 2:1. The aorta comes from the right ventricle and the pulmonary artery from the left ventricle. With modern newborn care, 1-year survival approaches 80%.

Typical presentation is a large, vigorous infant with cyanosis but little or no respiratory distress. There may be no murmur or a soft, systolic ejection murmur.

Chest x-ray study may be normal, but typically it reveals a very narrow upper mediastinal shadow ("egg on a stick" appearance).

There are no characteristic ECG findings but echocardiography is diagnostic and typical findings are branching of the anterior great vessel into the inominate, subclavian, and carotid vessels and branching of the posterior great vessel into the right and left pulmonary arteries.

Like echocardiography, cardiac catherization is diagnostic and often therapeutic. Urgent cardiac catherization with balloon septostomy and later Mustard or Senning procedures (transposing venous return via an intra-arterial baffle) or early arterial switch operation are the methods of treatment.

2. Tetralogy of Fallot (TOF).

TOF is characterized by 4 anomalies: pulmonary stenosis, ventricular septal defect, overriding aorta, and right ventricular hypertrophy (RVH). There is a slight male predominance. Cyanosis usually signifies complete or partial atresia of the right ventricular overflow tract or extremely severe pulmonary stenosis with hypoplastic pulmonary arteries. The degree of right ventricular outflow obstruction is inversely proportional to pulmonary blood

flow and directly proportional to the degree of cyanosis. TOF with absent pulmonary valve may present later in infancy due to poor feeding (due to very large pulmonary arteries causing esophageal compression).

At physical examination the patient is cyanotic with a systolic ejection murmur along the left sternal border. Loud murmurs are associated with more flow across the right ventricular outflow tract, and softer murmurs, with less flow.

Chest x-ray study shows a small, often "boot-shaped" heart, with decreased pulmonary vascular markings. A right aortic arch is seen in about 20% of these infants.

The echocardiogram may be normal or demonstrate right ventricular hypertrophy (RVH). The only sign of RVH may be an upright T wave in V4R or V1 after 72 hours of age.

Echocardiography is usually diagnostic, with the demonstration of an overriding aorta, ventricular septal defect (VSD), and small right ventricular outflow tract.
Pulmonary blood flow may be ductal-dependent with severe cyanosis and may respond to ductal dilation using prostaglandin E. This measure allows more flexibility for planning cardiac catherization and surgical correction. Surgery (shunting or total correction) may be palliative.

Acyanotic heart disease

Infants with acyanotic heart disease will achieve PaO_2 levels of over 100 mm Hg after breathing 100% inspired oxygen for 10-20 minutes.

The infant who is not cyanotic will present with either a heart murmur or symptoms of congestive heart failure.

Specific acyanotic heart disease abnormalities:
1. Ventricular septal defect (VSD) is the most common congenital heart abnormality, with equal sex distribution. Murmurs are not heard at birth but typically appear between 3 days and 3 weeks of age. Congestive heart failure is unusual before 4 weeks of age but may develop earlier in premature infants. Symptoms and physical findings vary with the age of the patient and the size of the defect. Spontaneous closure occurs in half. Surgical correction is reserved for large, symptomatic VSD only.
2. Atrial septal defect (ASD) is not an important cause of morbidity or mortality in infancy. Occasionally, congestive heart failure can occur in infancy but not usually in the neonatal period.
3. Endocardial cushion defects include ostium primum-type ASD with or without cleft mitral valve and atrioventricular (AV) canal. These defects are commonly associated with multiple congenital anomalies, especially Down's syndrome. If marked AV-valve insufficiency is present, the patient may present with congestive heart failure at birth or in the neonatal period.

Objectives

This study wants to establish the incidence of heart congenital malformations when compared to the other congenital malformations and their frequency according to some factors: risk, social background, sex and prematurity.

Material and method

The study is based on clinical, paraclinical examinations and imagistic explorations which were performed on premature new born hospitalized in the Neonatology and Health Care Clinic Timisoara between 2003 and 2005.

Results and discussions

Of the 72 studied with congenital malformations, 33 (45,8%) had heart congenital malformations. Of these 3 (9%) were cyanotic lesions (transpositions of the great arteries) and 30 (91%) – acyanotic lesions.

Regarding the social background 18 (54,5%) were from urban areas and 15 (45,5%) from the rural areas (fig 1).

The repartition of the cases according to sex showed that 16 (48,4%) were female and 17 (51,6%) were male (fig. 2).

According to the prematurity: 12 (36,3%) were 1st grade prematures, 12 (42,4%) – second grade prematures, 3 (9%) – 3rd grade prematures and 4 (12%) – 4th grade prematures (fig. 3).

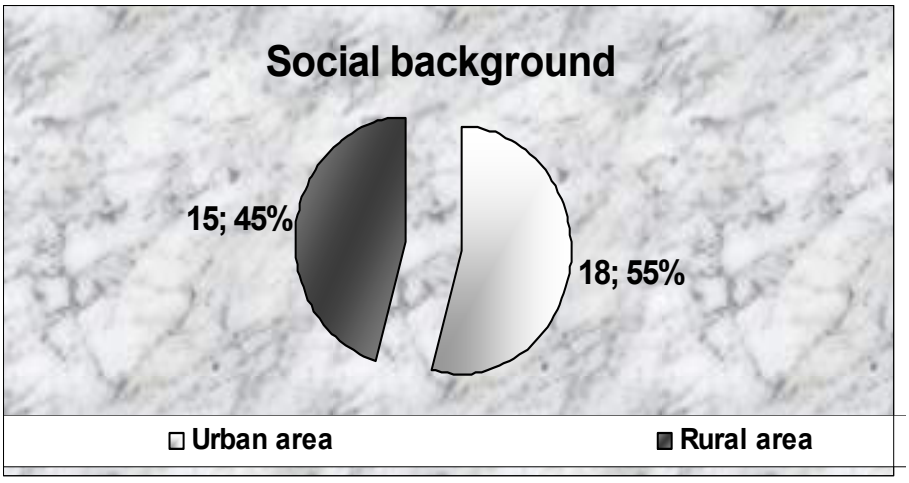

Fig. 1. Repartition of cases according to social background.

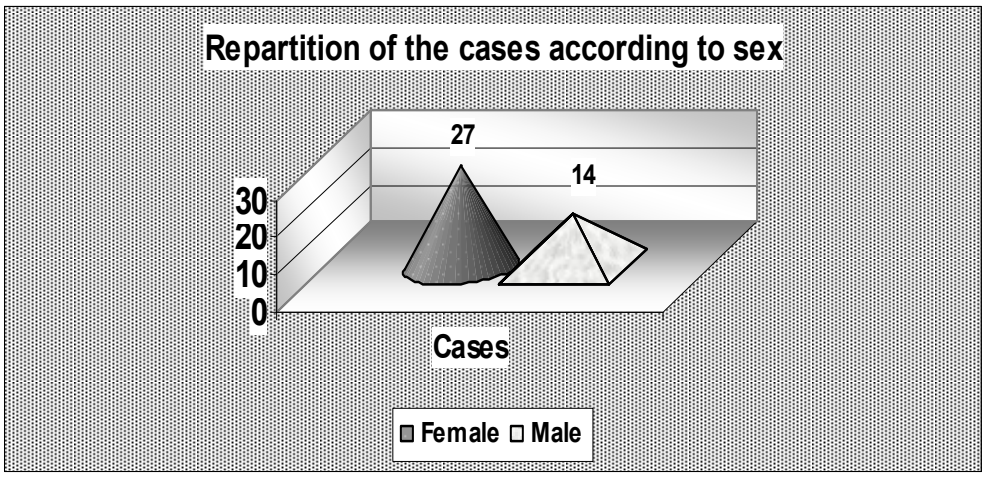

Fig. 2. Distribution of cases according to sex.

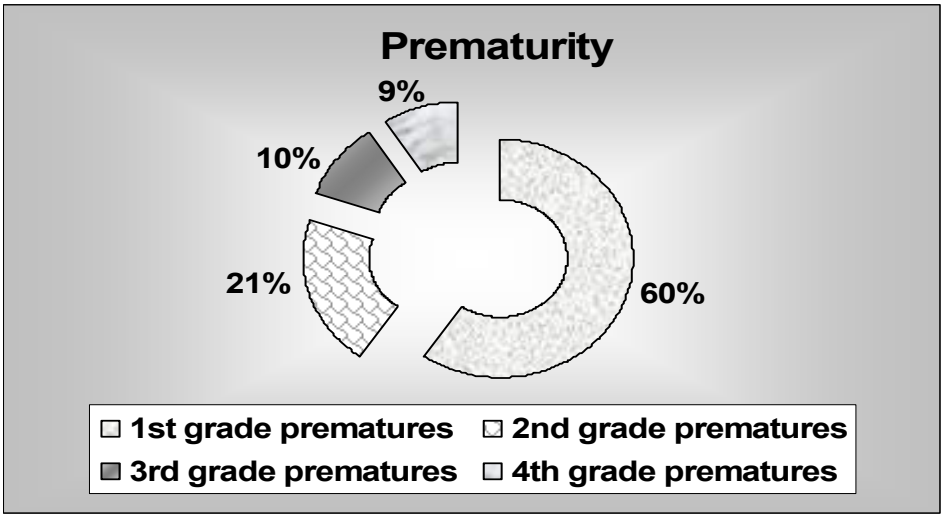

Fig. 3. Repartition of cases related to grade of prematurity.

Conclusions

1. The incidence of heart congenital malformations is high, representing 45,8% of the total congenital malformations.
2. The frequency of the acyanotic lesions is superior to the cyanotic lesions.
3. There is a slight predominance of the cases which come from the urban areas comparatively to those from the rural areas, possibly because of higher pollution.
4. Distribution according to sex is approximately equal.

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Correspondence to:
Daniela Iacob
D. Kiriac Street, No. 8, Ap. 9,
Timisoara 300487,
Romania
E-mail: danielariacob@yahoo.com