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Revista de Cercetare si Interventie sociala

ISSN: 1583-3410 (print), ISSN: 1584-5397 (electronic)

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Revista de cercetare și intervenție socială, 2018, vol. 63, pp. 379-388

The online version of this article can be found at:
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and
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REVISTA DE CERCETARE SI INTERVENTIE SOCIALA
is indexed by Clarivate Analytics (Web of Science) -
Social Sciences Citation Index
(Sociology and Social Work Domains)

Management of Neuropsychomotor and Economic Cost of Social Insertion of Children with Congenital Heart Malformations

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Abstract

Congenital Heart Defects (CHD) are a heterogeneous group of diseases present at birth, characterized by a structural heart defect. In Europe, their prevalence is of 8/1000 births. This study aims to review literature as to integrate neuropsychomotor, economic and psychological aspects in children with CHD. Therefore, the article presents the etiological factors of cardiac pathology, as well as the operative and post-operative etiological factors that may influence the neuropsychomotor and psychological development of a child with congenital heart defects, and financial costs involved for treatment and prevention.

Keywords: congenital heart malformations, child, psychosocial, economic implications.

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Introduction

Congenital Heart Defects (CHD) is a heterogeneous group of diseases characterized by a structural heart defect present at birth. Dolk, Loane, & Garne (2011) conducted in 16 European countries revealed the presence of CHD in 8 out of 1000 births. Moderate to severe CHD shown by Hoffman & Kaplan in 2002 based on 62 studies in different countries and continents (USA, Europe, Australia, China) is estimated to be of 6/1000 new-borns. Even after correcting heart surgery, paediatric patients may show residual anatomic abnormalities being prone to arrhythmias, pulmonary or systemic arterial hypertension, myocardial and coronary artery pathologies (Beers, Porter, & Jones 2009; Warnes *et al.*, 2001). About 5% of children have a chromosomal abnormality, (18, 13, 21 trisomy or Turner syndrome). Predisposing factors in the development of CHD are the maternal diseases (diabetes, rubella, collagen disease), certain toxic exposures (thalidomide, isotretinoin or alcohol), hypoxia (PCA is more common in populations living at high altitude), exposure of pregnant women to ionizing radiation or a combination of these factors. Generally, a specific cause cannot be identified. If we consider groups of patients with CHD who use public health services, we may observe a clear difference between patients with serious lesions, such as Tetralogy of Fallot, atrioventricular canal and patients with minor lesions, such as ventricular septal defect. Therefore, it is necessary to classify CHD in three groups of severity (*Table 1*).

Table 1. Classification of CHD according to severity

Classification	Types of CHD	Total incidence of CHD	Total incidence in new-born
CYANOGEN	Tetralogy of Fallot	3.5-8% (Schwartz, 2000)	3-6 / 10 000 (Longmore *et al.*, 2010)
	Transposition of great arteries	5-7% (Schwartz, 2000)	20.2 – 30 / 100 000 (Schwartz, 2000)
	Tricuspid atresia	1-3% (Beers, Porter, & Jones, 2009)	1/ 10 000 (Rao, 2013)
	Pulmonary artery atresia	0.7-3.1% (Charpie *et al.*, 2014)	4.5/ 100 000 (Charpie *et al.*, 2014)
	Hypoplastic left heart syndrome	1% (Beers, Porter, & Jones, 2009)	0.16 – 0.36 / 1 000 (Rao, 2013)
	Common arterial trunk	1-2% (Beers, Porter, & Jones, 2009)	0.1 (Rao & Turner, 2013)

NON-CYANOGEN	Atrial septal defects	6-10% (Beers, Porter, & Jones, 2009; Carr *et al.*,, 2014)	0.67 – 2.1 / 1 000 (Carr *et al.,* 2014)
	Ventricular septal defect	20-25% (Beers, Porter, & Jones, 2009)	1.5-2/ 1 000 (Schwartz, 2000)
	Atrioventricular canal	5% (Beers, Porter, & Jones, 2009)	0.24 – 0.31/ 1 000 (Brian, 2006)
	Patent ductus arteriosus	5-10% (Kim *et al.*, 2014)	0.02 – 0.06/ 1 000 (Kim *et al.*,, 2014)
OBSTRUCTIVE	Pulmonary stenosis	7-12% (Schwartz, 2000)	1 /2 000 (Beers, Porter, & Jones, 2009)
	Aortic stenosis	1% (Schneider & Singh, 2015; Krishnan & Singh, 2013)	8/10 000 (Schneider & Singh, 2015)
	Coarctation of the aorta	6-8% (Schwartz, 2000)	4/10 000 (Reller *et al.,* 2008)

Neuropsychomotor aspects in children with congenital heart defects

Etiological factors influencing the neurological integrity of paediatric patients with congenital heart defects are: length of hemodynamic and hematologic effects produced by the disease, hypoxic and ischemic lesions secondary to hypothermia used while performing the cardiopulmonary by-pass (Hickey, 1998). In patients with chromosomal abnormalities, especially with 21 trisomy and chromosome 22q11 deletion (Di George syndrome), the neurological development seems to be related to a genetic disease and not to cardiac defect (Daliento, Mapelli, & Volpe, 2006). Studies showed that cyanotic congenital heart diseases have an increased risk of stroke, especially during episodes of cyanosis or when blood viscosity is increased. Metabolic acidosis secondary to hypoxia accentuates the negative effects of ischemia on glial cells and cerebral vasculature by the action of free radicals on lipids and membrane proteins. Preoperatively, the clinical and electroencephalographic seizures and during prolonged activity on EEG were associated with increased risk of cognitive impairment.

Surgery performed early in the neonatal period or in the first trimester of life prevents cerebral complications. In patients with transposition of great arteries, the early performance of surgery prevents the onset of hypoxia and persistent

metabolic acidosis. In the study of Daliento, Mapelli, & Volpe (2006) it was shown that there is a risk of brain lesions even in patients with non-cyanotic congenital heart defects without severe heart failure. Surgical correction of CHD includes protection methods of brain structures, such as: cardiopulmonary bypass and hypothermia induction. Both methods can have neurological side effects caused by: risk of microembolism during bypass, difficulty in obtaining the optimal temperature to induce cardiopulmonary bypass.

Risk factors for brain lesions caused by hypothermia are: cardiopulmonary bypass, prolonged especially in younger patients (more than 45-50 minutes), rapid induction of hypothermia (under 20 minutes), severe hypothermia, hyperglycemia before cardiac arrest and during reperfusion, severe cyanosis and blood hyperviscosity before surgery, immediate postoperative low cardiac output, impaired cerebral blood flow secondary to autoregulation process deficiency.

Postoperative brain dysfunction is a serious complication of pediatric cardiac surgery that prolongs hospitalization; therefore, the early detection of postoperative neurological problems is important for establishing the treatment needed for early rehabilitation. Additionally, preoperative identification of children at risk requires new strategies to reduce the negative neurological impact after cardiac surgery. Specialized literature reports specific tests used to identify the degradation of cerebral function after cardiac surgery: increased levels of S-100 protein or neuron specific enolase in postoperative patient and changes in EEG. Clinically, the postoperative cerebral dysfunction includes the following symptoms: convulsions, paralysis, choreoathetosis, impaired cognitive or psychomotor development. In children with pre-existing neurological problems or with chromosomal abnormalities, the neurological worsening was considered to be present if the skills acquired by learning disappeared after surgery.

Trittenwein *et al.* (2003) report in a study that brain repercussions discovered after pediatric cardiovascular surgery were: cerebral infarction, cerebral bleeding, hydrocephalus and serious cerebral atrophy, diagnosed by CT, MRI or by performing necropsy. The same study represents the following important predictors for postoperative negative neurological impact: young age, complex cardiac defects, metabolic acidosis and increased serum levels of lactic acid. These predictors are plausible as in the first year of life; the accelerated growth of the brain makes it more susceptible to the adverse effects of hypoxia. Specifically, in the neonatal period, lactic acid is described as an important predictor of neurological dysfunction due to hypoxia. More complex congenital heart defects require a more elaborate surgery leading to a higher risk of intraoperative complications.

Generally, most lesions are located in the cortical region of the brain, especially, in the visual cortex and in the parietal lobe that may cause difficulty in integrating images in space and in developing organizational skills (Griffin, Elkin, & Smith, 2003).

Intellectually, most researchers agree that patients with CHD are at increased risk of intellectual impairment, the higher is the cardiac dysfunction, the more impaired is the cognitive function. Studies have shown that patients with cyanotic heart disease had lower IQ scores and learning difficulties. Noteworthy is the importance of differentiating between the effects produced by chronic disease in children and the development of cognitive impairment caused by CHD.

Wray & Sensky (1998) cognitively compared three groups of children: one group of children with cyanotic CHD, the second group of children with bone marrow transplant and a control group of healthy children. They demonstrated that children with cyanotic CHD had lower IQ than the others, with degradation in their reading skills and speed of response in 12 months after the first test, other groups remained constant. Some children with CHD have subtle neurological disorders that may be overlooked, although visual and fine motor perceptual disturbances are common in children with CHD. Such disorders may indicate visual and fine motor perceptual impairment or inability to process information correctly. Most studies reviewed by this study suggested that open heart surgery can produce neurological side effects. The study showed that the duration of cardiopulmonary bypass is negatively correlated with cognitive functions. The longer the child is connected to the bypass, the lower are IQ values and the child is more likely to develop mental and learning disabilities (Griffin, Elkin, & Smith, 2003).

Psychological Aspects in children with CHD

Psychologically, it is well acknowledged that most diseases impact the individual's well-being. Any chronic pathology diagnosed early which requires frequent medical assessment and therapeutic interventions may adversely affect the emotional balance and behavioral adjustment. This is applicable to chronic heart diseases, namely, CHD and especially serious and life-threatening diseases. Psychological implications play a significant role in the prognosis and evolution of chronic diseases (Johnson & Francis, 2005).

CHD diagnosed at birth may influence the mother-child relationship starting with infancy, can important period in psychological development of a human being; it may affect in the long run the mental health of children and teenagers. They may suffer from anxiety, depression, low self-esteem or impulsiveness. If not diagnosed and treated early, these emotional and behavioural disorders can lead to significant psychological and psychiatric morbidity; the prevalence of psychiatric disorders is 3-4 times higher in patients with neuro-cognitive impairment than in general population.

Complex mental and behavioural disorders especially in patients with CHD disorders have been noted, these are classified into two categories: externalizing (attention deficit disorder, aggression) and internalizing (anxiety, depression, somatisation) disorders (Marino *et al.*, 2012). In contrast, other studies have found

that chronological age and not the severity of congenital defects are significant risks in psychological development, chronological age, not the severity of congenital malformations of heart, noting that children and teenagers with CHD face higher risk of developing internalizing problems caused also by parental overprotection. Alternatively, brain and hormonal changes that occur during this period, combined with genetic vulnerabilities can enhance behavioural disorders (Karsdorp *et al.*, 2007). Neuropsychological differences are observed not only between children with CHD and healthy children and also between children with non-cyanogen and cyanogen CHD. Gupta *et al.* (1998) have objectified that patients with cyanotic congenital heart disease have an increased risk of developing depression, anxiety and behavioural problems

The type of medical and surgery treatment undergone by the child is important for his behavioral and psychological development. It has been found that: a high intensity therapy was linked with higher rates of behavioural problems (Janus & Goldberg, 1997). Children who underwent heart surgery have several behavioural disorders after surgery (Oates *et al.*, 1994). In contrast, the study by Wray & Sensky in 1998 found that children, who considered they to be shy preoperatively, postoperatively had an improved self-esteem. Other researchers noted a link between the total number of surgeries of a child and the induction of hypothermia with a higher rate of behavioral problems. Interiorizing disorders have been associated with an increased number of surgical procedures in which hypothermia was induced, small gestational age, low oxygen saturation and the age at surgery (Utens *et al.*, 1993).

The Economic costs of social insertion of children with CHD

The social impact is a major factor when the patient and parents become aware of the diagnosis with CHD. Most of these patients aspire to a normal social-economic and school life, an objective that can be achieved only if we foresee the problems and help patients solve them.

Prevention of infectious complications by vaccinations is essential for a favorable evolution. Influenza and pneumococcal vaccines are recommended for people with decompensated heart. There are no specific recommendations for patients with congenital heart disease but new recommendations for 2009 recommend vaccinations for patients with a a history of endocarditis. In what regards physical exercise, it is well-known that it has a major role in childhood not only in physical and motor but also in emotional, psychosocial and cognitive development (Bjarnason-Wehrens *et al.,* 2007). Physical inactivity in childhood is abnormal, children socialize through it and this can lead to socio-psychological problems. Exercise encourages children to accept teamwork, raises their self-esteem and teaches them what perseverance and respect for teammates and

opponents means (O'Brien *et al.*, 2012). Furthermore, sport teaches children with a competitive spirit, which helps them later in society on the one hand, in terms of performances, achieved, and on the other hand discipline on the labor market.

Additionally, sports helps to maintain normal weight because more than 25% of children with heart disease are overweight or obese. In this respect, it is important to give clear indications to parents about the type of physical activities that a child is allowed or not to do. If there are clear contraindications related to specific sports activities (severe PAH, Eisenmenger syndrome, single ventricle, coronary anomaly, severe Ebstein heart disease) parents should be given advice on specialized sports centres providing alternatives for sports activities aimed to help child's psychosomatic integration. New statistics show increased pregnancy in girls between 14-19. Pregnancy in teenagers with CHD is a special situation that put the young mother under high risk; therefore, well-organized family planning and discussions with the family and the teenager help in managing the problem. Estroprogestative contraception has increased efficiency, but these should be avoided when there is a risk of thromboembolism (cavopulmonary derivation, significant cyanosis).

Most patients can maintain pregnancy several times but it requires a multidisciplinary management (pediatric cardiologist, obstetrician, anaesthesiologist, neonatologist, genetician and haematologist). Some heart diseases can be aggravated by hemodynamic changes and hormonal impregnation duet o pregnancy. Pregnancy is contraindicated in severe PAH and Eisenmenger syndrome, maternal mortality is 50%. Pregnancy is a high risk in case of left ventricular dysfunction (LVEF <40%), dilated ascending aorta, desaturation <85% or mechanical valve. Clearly, the drugs prescribed for these patients are a risk for the fetus and should be stopped before the onset of pregnancy (ACE inhibitors and diuretics).

Supporting children with CHD in financial terms is also very important. Full insurance and compensation of medical interventions, development of specialized centres for managing such cases and provision of on-going information on the evolution and prognosis of children with CHD provide both to the child and family comfort and safety starting with the diagnosis.

Conclusions

The child with congenital heart defects presents a specific neurological, physical, psychological and cognitive development compared to a healthy child. Therefore, it is important to know and identify factors that influence neuropsychomotor development of a child with this pathology.

Even though surgery performed early in the neonatal period or in the first trimester of life prevents brain complications, especially in patients with transposition of

great arteries, preventing hypoxia and persistent metabolic acidosis, it has been demonstrated that surgery in itself can cause brain dysfunction.

Specialized literature identified the following predictors of adverse preoperative neurological impact: seizures discovered clinically and electroencephalographically, prolonged activity on EEG, young age, complex cardiac defects, metabolic acidosis and increased serum levels of lactic acid and postoperatively: long duration of cardiopulmonary bypass, increased levels of S-100 protein or neuro-specific enolase in patient serum, and postoperative altered EEG.

Economically and psychologically, most studies have shown the emotional and psychological impact of CHD diagnosis with mental and behavioural disorders, impact which translates into high financial costs, especially in patients with complex CHD classified into two categories: externalizing behaviour (attention deficit disorder, aggressiveness) and interiorizing behaviour (anxiety, depression, somatisation). Therefore, in order to reduce financial costs, it is important to diagnose and treat these psychological disorders early as they may lead to psychological and psychiatric morbidity, the prevalence of major psychiatric diseases being 3-4 times higher in patients with neuro-cognitive impairment than in general population.

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