Newborns with Suspected Congenital Heart Disease Demanding Emergency Transport: Seven Years Casuistic

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Abstract

Introduction: Congenital heart diseases are the most common congenital malformations in Portugal. Fetuses diagnosed *in utero* are referenced to and born in a tertiary care center. Newborns with suspected congenital heart disease missed in the prenatal screening program and born outside of a referral center that present with clinical severity are transported by the pediatric inter-hospital transport system. Our objective was to analyze seven years of the inter-hospital transport of newborns with suspected congenital heart disease in Portugal's northern region and study those who failed to be detected by the prenatal screening program.

Methods: Newborns with suspected congenital heart disease, transported between April 1, 2011 and March 30, 2018, were identified from the pediatric inter-hospital transport system records and individually reviewed in the national health service informatic system.

Results: The study includes 195 transports, corresponding to 144 patients. A mean of 28 transports per year were performed. Congenital heart disease was excluded in 46 patients. Of the 98 patients with congenital heart disease, 87 were born outside of the reference center. Transposition of the great arteries was the most common congenital heart disease transported (16/87, 18.4%) and aortic coarctation (14/87, 16.1%) was the second most common. Prenatal diagnosis was present in nine of these patients (10.3%).

Discussion: Transport number stability may translate the constant incidence of congenital heart diseases and/or reflect a non-improving prenatal diagnosis rate. Technical challenges in the transposition of great arteries prenatal diagnosis may explain the higher number of transports. It is necessary to guarantee the prenatal diagnosis of congenital heart diseases by intervening locally.

Keywords: Aortic Coarctation/epidemiology; Emergency Medical Services; Fetal Heart/diagnostic imaging; Heart Defects, Congenital; Infant, Newborn; Portugal; Prenatal Diagnosis; Transportation of Patients; Transposition of Great Vessels/epidemiology

Introduction

Congenital heart diseases (CHD) are the most common congenital malformation.[1-3] In Portugal, the prevalence was 86.5 per 10,000 births in 2014 and 2015,[4] which accounts for 800 children per year and 13/155 neonatal deaths in 2017.[5,6]

Fetal echocardiography is the gold standard for the prenatal diagnosis of CHD.[6] Second trimester ultrasound[7] identifies most of the suspicious fetuses who need further evaluation (non-identification of four proportioned chambers and the great arteries). Pregnancies with increased risk of CHD should be identified and referenced to a specialized center as early as possible. In addition to the suspicious cases detected in the second trimester ultrasound, all women with at least one risk factor for CHD should undergo a fetal echocardiography.[6]

Newborns with suspected CHD, not diagnosed in the prenatal period, who are born outside of a referral center and demonstrate clinical severity, need an appropriate transport to a hospital that can provide the best medical care.[8-10]

Portugal's northern region pediatric inter-hospital transport system[11] is responsible for the transport of CHD critically ill newborns for the reference center for congenital heart diseases in northern Portugal at Centro Hospitalar Universitário de São João (CHUSJ).

In this study, we analyzed seven years (from April 1 2011 to March 30, 2018) of the inter-hospital transport of newborns with suspected CHD of Portugal's northern region, calculated the prevalence of the CHD transported, and studied those which were not detected by the prenatal screening program and were not diagnosed *in utero*.

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Methods

The subjects included were newborns (until 28 days of life) who needed the emergency transport by the pediatric inter-hospital transport system to the reference center (CHUSJ) with confirmed or suspected CHD.

Potential patients were identified from the pediatric inter-hospital transport system records and individually reviewed in the medical, antenatal, and maternity records in the informatic system of the national health system (SClinico®).

All the patients transported between April 1, 2011 and March 30, 2018 with suspected or confirmed CHD were included. Six were excluded for a lack of transportation data and 20 were excluded for the absence of information on the definitive diagnosis.

Results

In the reviewed time period, 195 transports were considered, which account for 144 patients (Fig. 1).

From those 195 transports, 34 originated from CHUSJ and 161 originated from other 22 hospitals in Portugal's northern region. The destination of those transports was mainly CHUSJ (160/195). Twenty-seven transports had Lisbon surgical reference centers as the destination.

A mean of 28 (minimum 21, maximum 37) transports per year were performed. After 2016, no transports to Lisbon were identified. Within each year, the monthly distribution did not present a specific pattern.

Congenital heart diseases were excluded in 46 of these 144 patients. Therefore, 98 of the transported patients had CHD. Referring to these 98 patients, 61 patients were males (62.2%). Mean post-natal age at transport was 3.65 days (0-27 days) and 19/96 (19.4%) patients were born preterm. Mean weight at birth was 2,950.67 g (620-4,080 g) (Table 1).

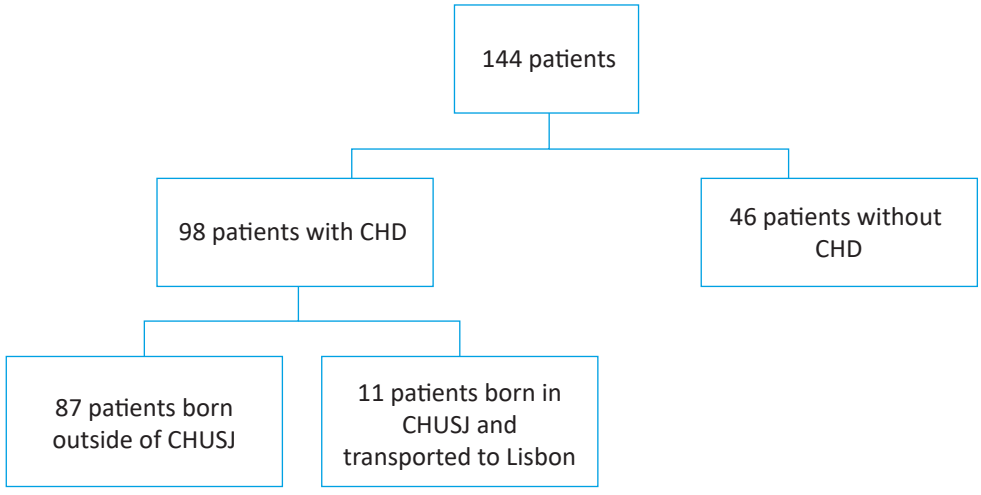
Presenting clinical signs were heart murmur (50/98, 51%), hypoxia (63/98, 64.3%), respiratory distress (35/98, 35.7%), hypotension or shock (15/98, 15.3%), and upper and lower limbs blood pressure differential (5/98, 5.1%) (Table 2).

Focusing on medical interventions to stabilize these CHD patients during transport, 28 (28.6%) were invasively ventilated and five (5.1%) patients were non-invasively ventilated. In 14 (14.3%) patients, inotropic drugs were given (dopamine and/or dobutamine), 40 (40.8%) patients were medicated with prostaglandin E1 (PGE1), 16 (16.3%) patients were sedated, and 12 (12.2%) patients were given antibiotics (ampicillin and gentamicin).

According to functional classification, CHD were grouped in left-sided obstructive CHD 29.6% (29/98), right-sided obstructive CHD 18.3% (18/98), parallel circulation 24.5% (24/98), left-to-right shunt 18.3% (18/98), and others 9.2% (9/98).

The most common CHD transported was the transposition of the great arteries (TGA) (24/98, 24.5%) and the second most transported CHD was aortic coarctation (14/98, 19.4%) (Table 3).

Prenatal diagnosis was present in 17 (17.3%) of those patients (6/24 transposition of the great arteries, 3/14 aortic coarctation, 2/6 tetralogy of Fallot, 2/4 hypoplastic left heart syndrome, 1/1 scimitar syndrome, 1/1 *truncus arteriosus*, 1/1 atrioventricular septal defect, and 1/1 right-sided obstructive CHD) (Table 3).



CHD - congenital heart defects; CHUSJ - Centro Hospitalar Universitário de São João.

Figure 1. Overview of the transported patients by the pediatric inter-hospital transport system of Portugal's northern region.

Table 1. Characteristics of neonates with confirmed congenital heart diseases transported by the pediatric inter-hospital transport system

Males, n (%)	61 (62.2)
Females, n (%)	37 (37.8)
Age at transport (days), mean ± standard deviation	3.65 ± 5.56
Preterm, n (%)	19 (19.4)
Term, n (%)	77 (78.6)
Weight at birth (g), mean ± standard deviation	2950.67 ± 715.7

Table 2. Presenting clinical signs of all the transported patients with confirmed congenital heart diseases

Clinical signs	**Transported patients (n = 98),** n (%)
Heart murmur	50 (51)
Hypoxia	63 (64.3)
Respiratory distress	35 (35.7)
Hypotension/shock	15 (15.3)
Blood pressure differential	5 (5.1)
Total	98 (100)

Among the 98 patients with confirmed CHD, 11 were born in CHUSJ and transported to another reference center in Lisbon. Hypoplastic left-heart syndrome was confirmed in two of these patients, transposition of the great arteries in eight, and *truncus arteriosus* in one patient. Prenatal diagnosis was present in eight of those patients.

Discussion

According to the literature, the specialized transport of critical children improves their outcome.[12,13] The transport of critical patients with CHD is discussed in this article.

The number of transports performed was superior to the number of patients because 51 patients needed more than one transport, either back to their hospital of origin or to a differentiated hospital for specific cardiac surgery in Lisbon. These transports from CHUSJ

Table 3. Congenital heart defects and prenatal diagnosis found in the transported patients with confirmed congenital heart diseases

		Transported patients (n = 98)	**Prenatal diagnosis**	
Functional classification	**Congenital heart diseases**	n (%)	**No**	**Yes**
Left-sided obstructive congenital heart diseases	Aortic arch hypoplasia	4 (4.1)	4	0
	Aortic atresia	1 (1.0)	1	0
	Aortic coarctation	14 (14.3)	11	3
	Aortic stenosis	4 (4.1)	4	0
	Hypoplastic left-heart syndrome	4 (4.1)	2	2
	Other	2 (2.0)	2	0
	Total	29 (29.6)	24	5
Right-sided obstructive congenital heart diseases	Pulmonary atresia	5 (5.1)	5	0
	Pulmonary stenosis	6 (6.1)	6	0
	TOF	6 (6.1)	4	2
	Other	1 (1.0)	0	1
	Total	18 (18.3)	15	3
Left-to-right shunt	AVSD	1 (1.0)	0	1
	IVC	7 (7.1)	7	0
	Multiple IVC + PDA	1 (1.0)	1	0
	TAPVC	8 (8.2)	8	0
	Other	1 (1.0)	1	0
	Total	18 (18.3)	17	1
Parallel circulation	TGA	24 (24.5)	18	6
Others	Biventricular hypertrophy	1 (1.0)	1	0
	Ebstein's anomaly	1 (1.0)	1	0
	Scimitar syndrome	1 (1.0)	0	1
	Truncus arteriosus	1 (1.0)	0	1
	Other	5 (5.2)	5	0
	Total	9 (9.2)	7	2
Total		98 (100.0)	81	17

AVSD - atrioventricular septal defect; IVC - interventricular communication; PDA - patent *ductus arteriosus*; TAPVC - total anomalous pulmonary venous connection; TGA - transposition of the great arteries; TOF - tetralogy of Fallot.
Others: complex cardiopathies with multiple associated cardiac defects.

to Lisbon (to Hospital de Santa Marta, Hospital de Santa Cruz and Hospital São Francisco Xavier) happened until 2015 because they were the centers that performed the correction of complex CHD. Once CHUSJ began to perform these surgeries, these extra transports ended.

The transport number stability throughout the seven years of this study may translate the constant incidence of CHD[14-17] and/or reflect a non-improving prenatal diagnosis rate.[1,8,14,18,19]

The predominance of male newborns transported found in our data is in accordance with male predominance in CHD,[20-22] with no specific explanation.

Antibiotic therapy was administered in 10.3% patients with CHD transferred to the tertiary center, probably reflecting a perinatal infectious risk.[23]

Referring to the clinical data, the need for ventilation, inotropic support, and PGE1 administration, reflects the severity of transported patients. This highlights the requirement of a specialized team to transport these newborns.

The number of cardiac anomalies detected on prenatal screening in this specific study population is obviously lower than the one found in the literature.[14,15,17] Our population only consists of critically ill patients born outside of the referral center that required emergency transport and not of all patients with CHD diagnosed in the northern region of Portugal.

Transposition of great arteries was the most frequent reason for transport, even excluding the transports to Lisbon. None of these patients with the transposition of great arteries had a prenatal diagnosis. The second most transported CHD was aortic coarctation, with three of the patients having a prenatal diagnosis. According to other authors, the most probable reason for this absence of prenatal diagnosis is the technical challenge in the transposition of great arteries diagnosis, since this CHD requires an outflow tract echographic view.[19,24] In Portugal, it is mandatory for the obstetrician to see the great vessels crossing in the fetus, therefore, in the view of this technical difficulty, the obstetrician should reassess or refer the pregnant woman to a fetal echocardiogram.[6] Reporting to aortic coarctation, the prenatal diagnosis is variable depending on the stenosis degree,[25-27] being easier to escape the prenatal diagnosis.

Every critical CHD patient should be born in a tertiary care center.[28] An ideally 100% prenatal diagnosis rate would imply a decrease in the need for transport. We know that some CHD, namely total anomalous pulmonary venous connection and mild to moderate aortic coarctation, are extremely difficult, if not impossible, to diagnose *in* utero.[29,30] These pathologies should be the only ones needing transport to a tertiary care center.

In conclusion, the stable frequency of CHD and the non-improvement of prenatal diagnosis is in accordance with other studies. Nonetheless, more efforts need to be made to improve the prenatal diagnosis of CHD, especially in those requiring outflow tract evaluation.

Future research for a better understanding of the reasons behind the failure in the prenatal diagnosis of the transposition of great arteries is urgent. Theoretically, there is no reason for this CHD not to be prenatally diagnosed and for these patients to be born outside of a tertiary care center.

WHAT THIS STUDY ADDS

- During the period of the seven years analyzed, 87 patients with congenital heart disease were transported to a reference center.
- Transposition of the great arteries was the most common heart condition, followed by aortic coarctation.
- The number of transports remained stable throughout the years, despite the efforts to improve prenatal diagnosis.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

Funding Sources

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Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data

The authors declare that they have followed the protocols of their work center on the publication of patient data.

Provenance and peer review

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Recém-Nascidos com Suspeita de Cardiopatia Congénita Exigindo Transporte de Emergência: Casuística de Sete Anos

Introdução: As cardiopatias congénitas são as malformações congénitas mais comuns em Portugal. Os fetos diagnosticados *in utero* são referenciados e nascem num centro de referência. Os recém-nascidos com suspeita de cardiopatia congénita que escapam ao programa de rastreio pré-natal e nascem fora de um centro de referência e que se apresentam com gravidade clínica são transportados pelo sistema de transporte inter-hospitalar pediátrico. O nosso objetivo é analisar sete anos de transporte inter-hospitalar de recém-nascidos com suspeita de cardiopatia congénita da zona norte de Portugal e estudar os que escapam ao programa de rastreio pré-natal.

Métodos: Recém-nascidos com suspeita de cardiopatia congénita, transportados entre 1 de abril de 2011 e 30 de março de 2018, foram identificados nos registos do sistema de transporte inter-hospitalar pediátrico e revistos individualmente no sistema informático do sistema nacional de saúde.

Resultados: Foram incluídos 195 transportes, correspondentes a 144 doentes. Foram realizados em média 28 transportes por ano. Foi excluída cardiopatia congénita em 46 doentes. Dos 98 doentes com cardiopatia congénita, 87 nasceram fora do centro de referência. A transposição das grandes artérias foi a cardiopatia congénita mais transportada (16/87,18,4%) e a segunda foi a coartação da aorta (14/87, 16,1%). Diagnóstico pré-natal estava presente em nove desses doentes (10.3%).

Discussão: O número constante de transportes pode traduzir a incidência constante das cardiopatias congénitas e/ou refletir uma não melhoria no diagnóstico pré-natal. As dificuldades técnicas no diagnóstico pré-natal da transposição das grandes artérias pode explicar o maior número de transportes. É necessário garantir o diagnóstico pré-natal das cardiopatias congénitas intervindo localmente.

Palavras-Chave: Cardiopatias Congénitas; Coartação Aórtica/epidemiologia; Coração Fetal/diagnóstico por imagem; Diagnóstico Pré-Natal; Portugal; Recém-Nascido; Serviços Médicos de Emergência; Transporte de Doentes; Transposição dos Grandes Vasos/epidemiologia