

Congenital Heart Defects

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Congenital heart defects are the most frequent congenital malformations. Severe congenital heart defects still represent one of the main causes of infant mortality. The risk factors associated with cardiac surgery and postoperative mortality are well known. Newborns with symptomatic congenital heart defects without an antenatal diagnosis, and who are delivered outside a referral center, need appropriate transport in order to receive the best medical care. In Northern Portugal, the pediatric interhospital transport system is responsible for the transport of critically ill newborns with congenital heart defects to the reference center at Centro Hospitalar Universitário de São João (CHUSJ). This issue of the Portuguese Journal of Pediatrics reports on the experience over the last seven years (1 April 2011 to 30 March 2018) with interhospital transport of newborns with suspected congenital heart defects in Northern Portugal.¹ Eighty-seven patients without an antenatal diagnosis were transported to CHUSJ (transposition of the great arteries was the most common defect, followed by aortic coarctation) and 11 (eight with prenatal diagnosis) were delivered to CHUSJ and then transported to another reference center in Lisbon (hypoplastic left-heart syndrome, transposition of the great arteries, and *truncus arteriosus*).¹ The number of congenital heart defects detected in prenatal screening, in this specific population, is lower than the one described in the literature,^{2,4} although the population only consists of critically ill patients born outside of the referral center that required emergency transport and not of all the patients with congenital heart defects diagnosed in Northern Portugal.

The number of transports remained stable throughout the years, despite the efforts to improve antenatal diagnosis. This report highlights three important aspects related to congenital heart defects¹:

- The absence of an antenatal diagnosis in several cases and, consequently, the risk of birth in a center without pediatric cardiology;
- The need for a 24/7 transfer team;
- The fact that some cases required a second transfer

to another center in Lisbon after stabilization at CHUSJ. Transports from CHUSJ to Lisbon (Hospital de Santa Marta, Hospital de Santa Cruz, and Hospital de São Francisco Xavier Hospitals) happened up until 2015 when CHUSJ began to carry out the correction of complex congenital heart defects and, since then, this aspect has already been solved.

Despite a decrease in the number of deaths due to congenital heart disease in the first year of life, the Portuguese Directorate-General of Health (DGS) identified the risk factors and women at risk for fetal congenital heart defects and determined within the scope of its technical-normative competences that:

- Pregnant women with at least one of the identified factors should be referred as early as possible to a high obstetric risk hospital clinic;
- If there is a suspicion of fetal heart disease, the pregnant woman should be sent directly to an obstetrics department that has the necessary resources, namely, support for prenatal cardiology and medical genetics;
- All initiatives aimed at improving the collaboration and coordination between primary health care, perinatal support hospitals, and differentiated perinatal support hospitals are encouraged and supported in order to ensure the correct and timely referral of pregnant women, circulation information, and training of human resources.⁵ However, efforts are still needed to ensure that an increasing number of heart diseases are correctly and timely diagnosed in the prenatal period. In this sense, it is necessary to improve the identification and guidance of women at risk in terms of primary health care, ensure the quality of the ultrasound exams of pregnancy, and the study of the fetal heart. Fetal echocardiography is the test of choice that allows for the prenatal diagnosis of a very large number of congenital heart diseases. The prenatal diagnosis of congenital heart defects is effective in reducing mortality as well as the morbidity that results from a late diagnosis. Every effort should be made in order to improve the early diagnosis of congenital heart defects as well as the timely guidance of pregnant women and neonates to referral centers.

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Conflicts of Interest

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