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ORIGINAL ARTICLE

Association between congenital heart defects and severe infections in children with Down syndrome☆

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KEYWORDS
Down syndrome;
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Abstract
Introduction: There is a high prevalence of congenital heart disease (CHD) in Down syndrome (DS) patients. Children with DS and CHD also present greater susceptibility to pulmonary infections than those without CHD.
Aim: To investigate the prevalence and types of CHD and their association with severe infections in children with DS in southern Brazil seen in a reference outpatient clinic.
Methods: Children aged between six and 48 months with a diagnosis of DS were included consecutively in the period May 2001 to May 2012, and the presence of CHD and severe infections (pneumonia and sepsis) was investigated, classified and analyzed.
Results: A total of 127 patients were included, of whom 89 (70.1%) had some type of CHD, 33 (37.7%) of them requiring surgical correction. Severe infections (pneumonia and sepsis) were seen in 23.6% and 5.5%, respectively. Of the cases of pneumonia, 70% had associated CHD (p=0.001) and of those with sepsis, 85% presented CHD (p=0.001).
Conclusions: Our study showed a high prevalence of CHD and its association with severe infections in children with DS seen in southern Brazil.

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PALAVRAS-CHAVE
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Associação entre cardiopatias congênitas e infecções graves em crianças com síndrome de Down

Resumo
Fundamento: Defeitos cardíacos congênitos (DCC) têm alta prevalência em pacientes com síndrome de Down (SD). Além disso, crianças com SD que possuem DCC são mais suscetíveis às infecções pulmonares do que aqueles que não possuem cardiopatia.
Objetivos: Investigar a prevalência, tipos de DCC e a sua associação com infecções graves em crianças com SD do sul do Brasil, atendidas em um ambulatório de referência.
Métodos: Durante o período de maio de 2011 a maio de 2012, foram incluídas consecutivamente no estudo crianças entre 6-48 meses de idade, diagnosticadas com SD nas quais foram investigadas, classificadas e analisadas as cardiopatias e infecções graves (sepse e pneumonia).
Resultados: Foram incluídos no estudo 127 pacientes. Desses, 89 (70,1%) possuíam algum tipo de cardiopatia, sendo necessária a correção cirúrgica em 33 (37,7%) deles. Com relação à presença de infecções graves, pneumonia e sepse foram diagnosticadas respectivamente em 23,6 e 5,5% dos casos. Dentre os casos de pneumonia, 70% das crianças apresentavam cardiopatia (p = 0,001) e nos casos de sepse em 85% eram cardiopatas (p = 0,001).
Conclusões: O presente estudo demonstrou alta prevalência de diferentes formas de DCC e a sua associação com infecções graves em crianças com SD atendidas no sul do Brasil.

Introduction

Down syndrome (DS) is the most common genetic cause of mental retardation worldwide, affecting 1/700 live births.[1] Among the various findings related to DS, a constant concern is the high prevalence of congenital heart disease (CHD), reported to affect 40–60% of children born with DS, depending on the population studied.[2–5] The most common is atrioventricular septal defect (AVSD) (30–60%), followed by ventricular septal defect (VSD) (around 30%); others include ostium secundum atrial septal defect (ASD) (around 10%), patent ductus arteriosus (PAD) and tetralogy of Fallot (TOF). Mitral valve prolapse may occur around the age of 20, with or without tricuspid valve prolapse and aortic regurgitation.[6] CHD remains a major cause of morbidity and mortality in the neonatal period,[7] due either to the heart defects themselves or to secondary factors, mainly associated with the compromised immune systems of these patients.[8] Children with DS and CHD are more susceptible to pulmonary infections than those without CHD.[9]

Given that DS is the most common chromosomal anomaly among newborns, that the life expectancy of these patients has increased and that early diagnosis of CHD is of the utmost importance, the present study set out to investigate the prevalence and types of CHD and their association with severe infections in children with DS in southern Brazil seen in a reference outpatient clinic.

Methods

The present study was approved by the ethics committee of Hospital de Clínicas da Universidade Federal do Paraná (HC-UFPR) and written informed consent was obtained from all participants. It was performed in the DS outpatient clinic of HC-UFPR in Curitiba, Paraná, during the period May 2011 to May 2012. Children aged between six and 48 months diagnosed with DS at birth based on clinical signs and confirmed by karyotype analysis were included consecutively. The investigation was based on a questionnaire completed by the family member accompanying the patient, together with review of the patient's medical records. Patients whose parents or guardians refused to participate were excluded from the study.

The CHD observed was classified according to Neonatologia – Pediatria Instituto da Criança HC/FMUSP 2011 using diagnostic methods in accordance with the recommended protocol (Avaliação cardiovascular do Neonato – Rev SOCERJ 2000). These included a detailed history, thorough physical examination, arterial blood gas analysis, chest X-ray, echocardiography and electrocardiogram.

The severe infections analyzed in the study were sepsis and pneumonia, which were classified according to the Brazilian guidelines for treatment of severe sepsis.[10] The information was recorded in a database, and analyzed using GraphPad Prism 4.0 statistical software.

Results

A total of 127 patients were included in the study, median age 18 months (6–48 months; mean 20.7±14.6 months), 37.8% (48/127) female and 62.2% (79/127) male. Mean birth weight was 2759±620 g (1080–4470 g) and mean length was 46.1±3.27 cm (34.5–53 cm). Mean maternal age was 32.9±0.67 years (17–49), and 48% (61/127) were aged over 35 and 24.4% (31/127) were aged over 40.

In accordance with Vaz et al.,[7] CHD was classified as follows: VSD, pulmonary atresia or stenosis, AVSD, TOF, ASD, coarctation of the aorta, and PDA. Of the 127 patients with DS, 89 (70.1%) had some form of CHD, of whom (52.8%) had more than one form (Table 1).