

# Odontological assistance to Down Syndrome patients having congenital heart malformations

*Assistência odontológica a pacientes com Síndrome de Down e portadores de cardiopatia congênita*

*Asistencia odontológica a pacientes con Síndrome de Down que tienen cardiopatías congénitas*

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## Abstract

In this study the authors present the frequency and types of congenital heart malformations (CHM) among Down Syndrome (DS) patients emphasizing the prevention of infectious endocarditis (IE) with appropriate antibiotic prophylaxis (ABP). Out of 390 DS patients, 312 (80%) were considered free from any CHM. 78 (20%) presented some CHM; from these 11,54% (n=9) have more than one CHM; ABP to prevent IE was recommended for 41,03% (n=32). Ventricular septal defect was the most frequent CHM (20,51%, n=16). Dentists must know about the patients' cardiologic diagnosis before a treatment that could cause bleeding, because they have to administer antibiotics to prevent IE. Although some CHM doesn't need ABP, according to the protocol of the American Heart Association, there are systemic conditions in DS that are relevant to the prescription of antibiotics.

**Descriptors:** Down Syndrome; Heart Defects, Congenital; Endocarditis, Bacterial ; Antibiotic Prophylaxis; Dentistry.

## Resumo

Neste estudo, os autores apresentam a frequência e os tipos de cardiopatias congênitas (CHM), entre pacientes com Síndrome de Down (DS), enfatizando a prevenção de endocardite infecciosa (IE) com a profilaxia com antibióticos apropriados (ABP). Dentre 390 pacientes com SD, 312 (80%) foram considerados livres de qualquer CHM. 78 (20%) apresentaram algum tipo de CHM; destes 11,54% (n = 9) têm mais do que um tipo de CHM; ABP para evitar IE foi recomendado para 41,03% (n = 32). Defeito septal ventricular foi a mais frequente CHM (20,51%, n = 16). Cirurgiões-Dentistas devem saber sobre o diagnóstico cardiológico dos pacientes antes de um tratamento que possa causar sangramento já que essa situações exigem antibioticoterapia preventiva. Embora alguns tipos de CHM não exijam ABP, de acordo com o protocolo da American Heart Association, há condições sistêmicas no DS que são relevantes para a prescrição de antibióticos.

**Descritores:** Síndrome de Down; Cardiopatias Congênitas; Endocardite Bacteriana; Antibioticoprofilaxia; Odontologia.

## Resumen

En este estudio, los autores presentan la frecuencia y el tipo de cardiopatía congénita (CHM), entre los pacientes con Síndrome de Down (DS), haciendo hincapié en la prevención de la endocarditis infecciosa (IE) con la profilaxis con antibióticos apropiados (ABP). Entre 390 pacientes con SD, 312 (80%) fueron considerados libres de CHM. 78 (20%) tenían algún tipo de mecanismo de facilitación; los 11,54% (n = 9) tener más de un tipo de CHM; BPA para evitar que IE se recomendó a 41.03% (n = 32). Defecto septal ventricular fue el CHM más frecuente (20,51%, n = 16). Los dentistas deben saber sobre el diagnóstico cardiológico de los pacientes antes del tratamiento que pueden causar sangrado, ya que las situaciones que requieren tratamiento preventivo con antibióticos. Aunque algunos tipos de CHM no requieren BPA, de acuerdo con el protocolo de la American Heart Association, existen condiciones sistémicas en DS que son relevantes para la prescripción de antibióticos

**Descriptor:** Síndrome de Down; Cardiopatías Congénitas; Endocarditis Bacteriana; Profilaxis Antibiótica; Odontología.

## INTRODUCTION

Down Syndrome (DS) is an autosomal chromosomal anomaly which results from trisomy of all or part of chromosomal 21. It is the single most common genetic cause of mental retardation affecting approximately 1 in 700 live births<sup>1</sup>.

CHM in DS patients occur in about 8 to 10 cases in 1,000 births. In average, 20% of these patients have more than one type of CHM<sup>2</sup>.

Many of the syndromes related to chromosomal abnormalities and mental retardation are frequently associated with CHM. Among them DS is the highest in number of cases<sup>2</sup>. This is the chromosomal disturbance more common and well-known. It's very common in mothers after 35 years old. Considering the behavior, DS patients, on average, tolerate dental office procedures more frequently than other mental deficiency patients<sup>3-5</sup>.

There are possibilities of occurrence of miscarriages during a gestation period of a Down Syndrome infant<sup>4</sup>. The incidence of CHM is high in these patients and constitutes a high rate of morbidity and mortality before and after birth.

Atrioventricular canal as well as septal defects and tetralogy of Fallot are the most common CHM in DS children. In average, 30 to 40% of the patients present isolated CHM<sup>2,4-6</sup>. However, more than 30% present multiple CHM<sup>6</sup>.

In the oral cavity there is a large number of microorganisms and any mouth hygiene or dental procedures may cause bleeding and bacteriemia<sup>7</sup>.

The agents more frequently found in the IE cases are: alpha-hemolytic streptococci (50 to 60%), staphylococci (10 to 20%), enterococci (15%), fungi and gram-negative bacteria (15%). Several different microorganisms can attack the heart lesion simultaneously<sup>8,9</sup>.

Bacteriemia doesn't pose as a high risk for a healthy patient, due to the immunological system being capable of combating the microorganisms. The same doesn't happen with patients who have CHM or

acquired heart malformations (AHM), orthopedic or valve prostheses, transplanted organs and/or immunodepression. In these cases there is a very high risk of IE<sup>7,10</sup>.

*American Heart Association* recommends ABP to prevent IE in certain medical conditions such as users of valve prostheses, most of CHM, AHM, previous bacterial endocarditis, among others. Possible antibiotics to be used are: amoxicillin, ampicillin, clindamicin, cefalexina, cefadroxil, azitromicin, claritromicin and cefazolin. Dosage and length of administration should follow recommendation<sup>11,12</sup>.

Individuals having the risk of developing IE should maintain good mouth hygiene as much as possible, reducing the proliferation of bacteria. Poor mouth hygiene, periodontal infections or ulcers caused by bad adapted prostheses can also develop bacteriemia, independent of having any dental procedure<sup>11,13</sup>.

The success of dental treatment can be obtained when the professional knows psychological status and general health of your patient. Therefore, the dentist can carry out the procedures safely<sup>14</sup>.

All the DS patients evaluated in this research were assisted at CAOÉ by a multi-professional team. Most of them, live in rural communities and belong to a low social, economic and cultural level. These factors contribute for the no accomplishment of pre-natal care of their mothers. Besides that these mothers were usually old for occasion of their gestations, what increases the possibility of them generate children with DS. The goal of this research was to present the most frequent types of CHM among DS patients emphasizing the prevention of IE with appropriate ABP.

## MATERIAL AND METHODS

From a list of patients assisted at CAOÉ, from 1985 to 2001, we detected 390 with DS to perform this study. Diagnosis of DS was established from clinical evidences (signs and symptoms) and/or by cytogenetic

exam. Also the Cardiologist of CAOE evaluated them by a clinical check up and complementary exams, like electrocardiogram (ECG) or others if necessary.

Data regarding CHM contained in their clinical records were filed into the computer for a database, through the EpiInfo® 3.4 program (Centers for Disease Control and Prevention – CDC) for making tables and graphics.

## RESULTS

Out of the 390 patients, 312 (80%) were considered free from any CHM from birth or after a successful surgical correction. Only 78 of them (20%) presented some CHM.

The patients was separated in 8 groups: ASD (atrial septal defect), VSD (ventricular septal defect), PDA (patent ductus arteriosus), OAVC (*ostium atrium ventriculorum communis*), IHM (innocent heart murmur), MVP (mitral valve prolapse without regurgitation), AP (associated pathologies) and OTHERS.

The patients ranged from 0 to 50 years of age. The results are detailed in the tables 1 and 2 and figure 1.

**Table 1.** Frequency of Down Syndrome patients presenting or not CHM.

Heart Conditions	Men	%	Women	%	TOTAL	%
CHM	46	11.79	32	8.21	78	20
Healthy Heart	170	43.59	142	36.41	312	80
<b>TOTAL</b>	216	55.8	175	44.62	390	100.0

Group OTHERS: pulmonary stenosis (PS), n=2; aortic valve insufficiency (AVI), n=2; tricuspid valve insufficiency (TVI), n=1; mitral valve insufficiency (MVI), n=2 (1 had also OAVC, partially corrected and with residual heart murmur). Group AP: VSD + PS, n=1; VSD + MVI, n=1; VSD + subaortic stenosis, n=1; Tetralogy of Fallot (TOF) + Valvar and subvalvar pulmonary stenosis, n=1; MVI + TVI, n=1; MVP + MVI, n=1; MVP + AVI, n=1; DSV + DSA (residual heart murmur after surgery), n=1; DSV + PDA, n=1 (residual heart murmur after surgery).

## DISCUSSION

An innocent heart murmur, despite being the most frequent heart alteration found in our patients is not a CHM and therefore, the administration of ABP is not recommended. According Rothwell et al.<sup>13</sup> we can consider the group MPV (n=9) to be in the same situation because the pathologies are isolated without regurgitation.

We indicate ABP for all patients in the groups ASD (n=4), VSD (n=11), PDA (n=2) and OAVC (n=3), but it is not recommended by the *American Heart Association*<sup>11</sup> in the cases of ASD. The CAOE clinical team indicates it due to fragile immunity, frequent infections and comorbidities besides malnutrition due to poor socioeconomic condition. DS patients have abnormal immunological system that predisposes them to the serious infections.

**Table 2.** Frequency of each CHM (n=78).

CHM	MEN	% of total sample (n=390)	% of CHM sample (n=78)	WOMEN	% of total sample (n=390)	% of CHM sample (n=78)
ASD	3	0,77%	3,85%	1	0,26%	1,28%
VSD	5	1,28%	6,41%	6	1,54%	7,69%
PDA	1	0,26%	1,28%	1	0,26%	1,28%
OAVC	2	0,51%	2,56%	1	0,26%	1,28%
IHM	19	4,87%	24,36%	14	3,59%	17,95%
MPV	4	1,03%	5,13%	5	1,28%	6,41%
OTHERS	6	1,54%	7,69%	1	0,26%	1,28%
AP	5	1,28%	6,41%	4	1,03%	5,13%
<b>TOTAL</b>	45			33		

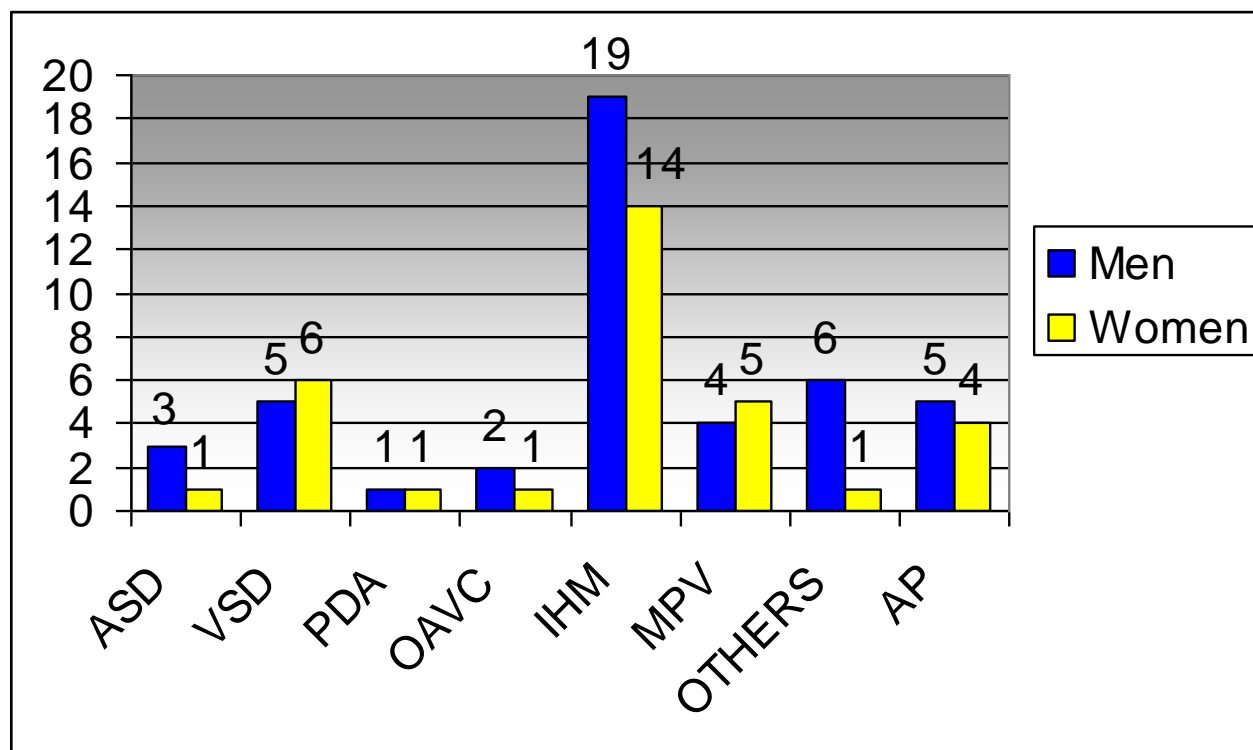


Figure 1. Graphic of congenital heart malformations found in 78 patients, according to the gender.

Besides, they have a risk from 10 to 20 times bigger to develop acute leukemia<sup>15</sup>. The nutritional lack causes illnesses that attack 40% of the population human (two billion people). Of these, hundreds of millions acquire irreversible sequels. There is a synergic relation between systemic pathology, nutritional deficiencies and infectious processes. The malnutrition compromises the coetaneous barriers and mucous, it diminishes the immunological defenses; the infections increase the necessity of nutrients, due to intensive protein catabolism. This harm the digestion, absorption (anorexy, vomits) and excretion of nutrients<sup>16</sup>.

The ABP is indicated in the group OTHERS (n=7) to 1 patient with MVI, 1 with AVI and 1 with TVI (high risk); it's not indicated to 1 with MVI, 1 with AVI and the PS patients (low risk). The risk was evaluated by clinical features.

The presence of residual heart murmur always does not mean that the surgical correction has not been a success<sup>17</sup>. However, residual heart murmur can reveal a CHM partially corrected with potential risk for the development of IE.

The ABP is indicated to all patients in the group

AP. The remaining individuals having CHM don't need ABP because their pathologies have been considered physiologically innocuous. Ventricular Septal Defect was the most frequent pathology (20,51%, n=16), to which the ABP is recommended by *American Heart Association*<sup>12</sup>.

In accordance to Kallen<sup>18</sup> 40% have CHM, responsible for the majority of the deaths in infancy. In our study this evidence was small. This difference creates hypotheses regarding the level of assistance to the health, methodologies of the studies, epidemiology and, also, genetic and environmental factors between the countries or regions of the samples.

Alabdulgader<sup>19</sup> mentions the determination of the real occurrence of the CHM requires accurate diagnosis. The author also emphasize that the modern diagnosis techniques provide higher incidences and that regional differences are observed in the occurrence of several heart lesions. We can consider there is a possibility that lack of more modern and accurate diagnosis methods have influenced our results. But we know about the need of making public our reality and shortcomings. This is a specific sample of patients from CAO. Considering the limits of this clinical



study, although that lack can bring a statistical meaning, this doesn't affect our safety for the recommendation of ABP. Furthermore, lack of epidemical data, mainly in Brazil, probably hinders any prospective study about CHM. Besides that, absence of evident clinical manifestations that happen in several of those CHM hinders even more these studies. When it refers to special care patients with CHM, the situation becomes more critical because little is known about this subject.

Using the fetal echocardiography, Paladini et al.<sup>20</sup> observed that 56% of 41 DS fetuses had some CHM, and the most common (44%) was Atrioventricular Septal Defect. The authors state that there are failings in diagnosing Ventricular Septal Defect during prenatal life, because it would be detected accurately only after birth. They stated that babies having CHM have shorter life expectancy than those free of CHM.

Moller and Anderson<sup>21</sup> states in a study carried out with 1,000 children having heart malformations that 33% presented Ventricular Septal Defect, 10% PDA and 10% PS.

Hijji et al.<sup>17</sup> in a study about life expectancy and social adaptation in DS patients showed that the survival until 24 years of age was 92.2% in patients free from CHM and 74.6% in patients with it. From these 74.6% that underwent corrective surgeries there was a percentage of survival of 87.8%. Among those that were not operated the percentage of survival was 41.4%. The most common CHM in that study was VSD. The authors recommended that DS children should go to a cardiologist to be evaluated and, if necessary to be submitted to a surgery in the first months of their life.

It's very important an interaction between doctors and dentists, because the pediatrician follows the children from birth offering them full assistance<sup>22</sup>.

In this study the relationship between survival and age shows a similarity with the mentioned studies. The average age of patients with a healthy heart was

14,78 years old and the ones with CHM and need of ABP was 12,82 years old.

Alabdulgader<sup>19</sup> in his study with 740 children pointed that VSD was the most common lesion and the DS was the only significant etiological factor associated to the occurrence of the CHM.

To establish criteria for dental procedures in DS, current clinical and updated scientific knowledge must be gathered through careful evaluation of each patient. This way, recommendations from *American Heart Association*<sup>11,12</sup> for the ABP to prevent IE are respected and applied by the professional team at CAO. However, the decision of the clinical team is sovereign in the evaluation of each patient. The criteria to carry out preventive procedures are: a) the kind of assistance: elective or urgent; b) knowledge of the cardiologic and systemic conditions. Every dental procedure that causes bleeding is, whenever possible, postponed until a definitive cardiologic diagnosis is obtained. Dental treatment should be accomplished in agreement with the diagnosis. However, when there is an emergency/urgency without the cardiologist's evaluation the ABP must be done.

Carmona et al.<sup>23</sup> found 12 cases of IE of oral origin: six were associated with oral infections and six with previous dental procedures. Besides, there were cases of IE patients with healthy heart. They concluded that is need better oral hygiene and improvement dental care in both 'at risk' and healthy patients. It's impossible to discriminate between those patients 'at risk' and those where transient bacteriemia poses as a risk of IE.

Studies have been comparing the magnitude of the auto-induced bacteriemias, caused by the patients themselves, and those induced by dentists. According to Pallasch<sup>24</sup>, "Royal College of Physicians of London" and "British Cardiac Society" less than 4% of the cases of IE can be associated with dental care procedures. Dajani et al.<sup>11</sup> also share a point of view that the bacteriemia produced due to daily procedures of the patient's buccal hygiene is more dangerous than that

produced by dental care procedures. Therefore, it's necessary to maintain good oral hygiene in the patients with risk of IE. The responsibility of the cases of IE involves patient, their relatives and health professionals, who must be always updated on technical and scientific knowledge<sup>25</sup>.

Mansur et al.<sup>26</sup> applied a questionnaire to 130 dentists and dentistry students to evaluate their knowledge on fundamental points that involve the aetiopathogenesis of IE. The preoccupying results showed that 23.8% didn't ask about the presence of heart murmur or rheumatic disease in their patients and 40.7% didn't prescribe ABP to the CHM. This data should be taken into consideration to make a change in the professional behavior.

The patient with CHM requests special care from the dentists, who should try to exchange information with the patient's doctor or a cardiologist. Like that, the dental treatment can to be carried out safely and successfully<sup>9,13,22,26</sup>.

## CONCLUSION

We concluded that:

1. 20% of all DS patients had some CHM;
2. From these patients:
  - The ABP to prevent IE is recommended for 41,03%;
  - 11,54% of them have more than one CHM;
  - VSD was the most frequent CHM (20,51%, n=16)
3. Dentists should know about the patient's cardiologic diagnosis before starting the dental treatment;
4. Although some CHM doesn't justify prescription of ABP, there are systemic conditions in DS patients that are relevant to the prescription.

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### **CONFLITO DE INTERESSES**

Os autores declaram não haver conflitos de interesse.

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