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(RESEARCH ARTICLE)

# Epidemio-clinical and echographic aspects of the atrio -ventricular canal in Madagascar

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## Abstract

**Introduction:** Atrioventricular canal defect (AVCD) is a congenital heart malformation characterized by a variable degree of atrial and ventricular septal defect [1] . Anatomically, there are partial, complete and intermediate forms [2] . This first Malagasy study was designed to describe its epidemio -clinical aspects in children with AVCD and to determine the factors influencing the prognosis but also to serve as a database of AVCD in Madagascar.

**Method:** This was a retrospective, descriptive and analytical study over a period of 20 years on 98 patients with AVCD seen at the *Soavinandriana Hospital Center (CENHOSOA)* Antananarivo.

**Results:** We found that AVCD represented 4.70% of congenital heart diseases observed with a mean age of discovery of 29.24 months and a male predominance (sex ratio of 1.61). The complete form represented 84.54% of cases. Trisomy-21 was found in 46.39% of patients. Atrial septal defect (ASD) (12.37%) and ventricular septal defect (VSD) (8.25%) were the main associated cardiac malformations followed by transposition of the great vessels (8.25%). Signs of heart failure were present in 27.83% of cases and 34.02% had pulmonary hypertension (PH) of which 11.34% had Eisenmenger syndrome. In this study, age of discovery more than 12 months was associated with the onset of PH and Eisenmenger syndrome (p = 0.03 and 0.01). Non-trisomy patients tend to present with heart failure and PH (p = 0.02 and 0.01).

**Conclusion:** Cardiovascular exploration is required in all infants with Down syndrome and those who present signs of CAV. Delayed diagnosis remains the main factor in poor prognosis of CAV in Madagascar.

**Keywords:** Atrioventricular Canal; Echocardiography; Epidemiology; Prognosis; Screenig

## 1. Introduction

Robert Anderson described in 1982 the different congenital heart diseases, in particular atrioventricular communication ( AVCD) formerly called atrioventricular canal malformation or endocardial cushion defect [1] . AVCD is a congenital heart defect characterized by a variable degree of atrial and ventricular septal defect [2] . Anatomically,

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there is the partial form characterized by an inter - atrioventricular communication with ostium primum and two separate orifices of the atrioventricular valves with a cleft of the anteromedial leaflet of the mitral valve. The complete AVCD includes an inter-atrioventricular communication with ostium primum, a common atrioventricular valve and a confluent posterior inter-ventricular communication located in the inlet portion of the ventricular septum. The intermediate form presents a restrictive inter-ventricular communication associated with the anatomical features of the partial AVCD [3] . AVCD is quite common is often associated with extracardiac malformations and has a strong genetic association especially with trisomy 21 [3] .

## 2. Method

This is a retrospective, descriptive and analytical study over a period of 20 years between 1997 and 2017. All patients who consulted the "*Chaine de l'espoir*" association at the *Soavinandriana Hospital Center (CENHOSOA)* Antananarivo were included in the initial database and were treated. All children diagnosed as AVCD on cardiac echo-Doppler were included. We studied sociodemographic data, associated malformations, clinical manifestations and echocardiographic data. Data were collected on Excel 2013 software, processed and analyzed on Epi Info 7.2.2.16.

## 3. Results

During the study period, 2021 children were affected by congenital heart disease. Among these children, 98 had AVCD representing 4.70% of congenital heart diseases. One file was excluded for lack of information and the remaining 97 were retained. The male gender was predominant with a sex ratio of 1.61. The average age of discovery was 29.24 months with a peak between 0 and 6 months. Trisomy 21 was present in 46.39% of cases. Cardiovascular assessment for trisomy 21 was the most common reason for consultation (38.14%) followed by heart murmur (32.99%). On clinical examination, 69.07% of children presented with a heart murmur and 24.74% with a burst of pulmonary S2. A picture of heart failure was observed in 27.84% of children. Complete AVCD represented 84.54% of cases, partial AVCD 14.42% and the intermediate form 1.03% of cases *(Figure 1).*

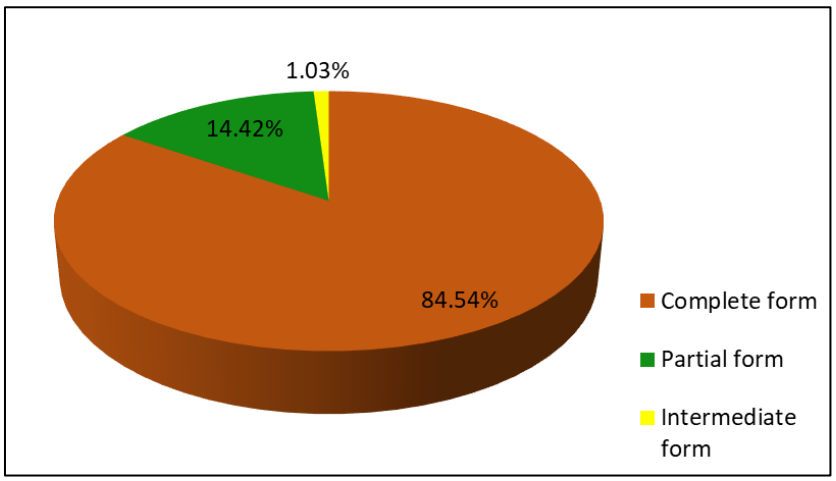


**Figure 1** Atrioventricular communication shape according to echocardiographic phenotype

The added CIA ostium secondum was the most frequent associated cardiac malformation (12.37% of cases) followed by transpositions of the great vessels (8.25%) and additional interventricular communication (IVC) (8.25%) *(Figure 2).*

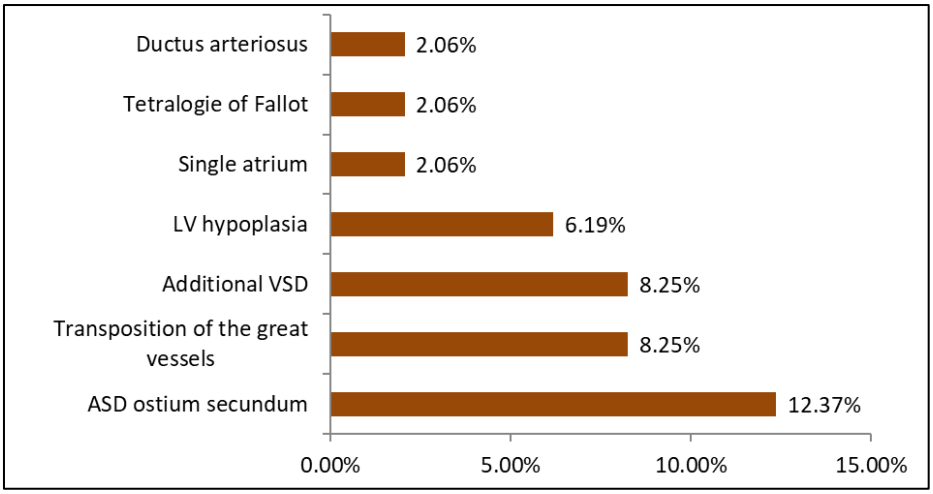


**Figure 2** Heart malformation associated with atrioventricular communication. LV, left ventricle; VSD, ventricularseptal defect; ASD, atrial septal defect

PH was present in 34.04% of children and 11.34% had Eisenmenger syndrome. Only 14.43% of these children had received surgical treatment who were all non-trisomic. There was a significant association on trisomy 21 and the complete phenotype of AVCD *(p = 0.01).* After analysis of the children's age, the age of discovery greater than 12 months was also associated with the presence of pulmonary hypertension on cardiac echo-Doppler *(p = 0.03).*

## 4. Discussion

AVCD represents an important entity of congenital heart disease but remains behind ASD, VSD, pulmonary stenosis, tetralogy of Fallot and transposition of the great arteries in frequency worldwide [4] . AVCD represented 4.70% of congenital heart disease in this study. In the Congolese study of *Nika et al*, CAV represented only 1.80% of congenital heart disease far behind tetralogy of Fallot , ASD and patent ductus arteriosus (PDA) [5] . A worldwide study described by *Liu et al* suggested a prevalence of 3.9% of AVCD among all congenital heart disease [4] .

There was a male predominance of children with AVCD in our study with a sex ratio of 1.60. The Santoro study in Italy and the *Fudge et al* cohort in the USA also found this male predominance of AVCD [6,7] . In the literature, trisomy 21 is described as more frequent in male subjects, on the other hand, congenital heart diseases associated with trisomy 21 including AVCD are more frequent in female subjects [6] .

There was a significant difference in the mean age at diagnosis in this study compared to those described in more developed countries. In the United States, in the studies of Fudge et al, and *Geva et al*, the mean age of discovery was 1.27 months and 0.87 months respectively [7,8] . In the Congolese study of *Nika et al* based on databases from the NGO *"Chaine de l'Espoir"* in Congo, the mean age at diagnosis was 51.4 +/- 50.4 months [5] . This diagnostic delay is mainly due to the inadequacy of reference centers for diagnosis in developing countries and the lack of knowledge of the population of the initial warning signs of AVCD. The age of children has a major influence on the surgical management and prognosis of children with congenital heart disease including AVCD [9] . Prenatal diagnosis is currently possible by echocardiography, often influencing the family's decision on whether to continue the pregnancy in developed countries, yet can help to provide early management at birth and a better prognosis [10] .

AVCD is one of the congenital heart diseases of people with Down syndrome [6,7] . Down syndrome is often associated with heart malformations, the most common of which are AVCD, ASD, VSD and tetralogy of Fallot. [11] . In this case series, 46.39% of children had Down syndrome. These results were rather similar to those in developed countries; in the study by *Huggon et al* in the United Kingdom and in the larger study by *Christensen et al* in Europe, Down syndrome represented 39% and 49% of patients with AVCD, respectively [9,12] . In the Italian study by *Santuro et al*, AVCD represented 39.6% of congenital heart disease associated with Down syndrome, just behind VSD at 45.5% of cases [6] .

AVCD is rarely isolated but often associated with other cardiac and extracardiac malformations within the framework of syndromic malformations [8,12] . In our study, ASD ostium secundum was the most common associated cardiac malformation, followed by transpositions of the great vessels and additional VSD.

The complete form of CAV was predominant in our series, representing 84.54% of cases. These results were similar to those of the study by *Geva et al* in the USA (68.40% of AVCD) and those of *Olariu et al*. in the United Kingdom (79.04% of AVCD) [8,13] . The complete form is the most frequent form of AVCD in both Down syndrome and non-Down syndrome patients [9] .

On the evolution and complications of AVCD, 11.34% of our patients had presented Eisenmenger syndrome. Eisenmenger syndrome is described as the elevation of pulmonary arterial pressure at the systemic level caused by increased pulmonary vascular resistance with inversion of the shunt through a cardiac defect linked to a cardiac malformation [14] . It is the ultimate evolution of the various congenital heart diseases not treated in time [14] . In developed countries, Eisenmenger syndrome is currently rare with the different diagnostic means and early management of children with congenital heart disease. Eisenmenger syndrome was present in 1.61% of children with AVCD in the study of *Olariu et al* in Romania [13] . A clinical and echocardiographic evaluation of Down syndrome patients should be systematic in the first six weeks of life to detect an associated cardiac malformation. Surgical management should be performed within 3 to 6 months of the child's life to avoid the onset of irreversible pulmonary arterial hypertension [9] .

Regarding surgical treatment, only 14.43% of AVCD were operated on in this study, all of which were non-trisomic patients. According to *Fudge et al*, there is no significant difference in the outcome of patients with trisomic and non-trisomic patients with surgical management of AVCD, but post-operative complications are more frequent in trisomic patients [7] .

## 5. Conclusion

AVCD is one of the main congenital heart diseases. It is one of the heart diseases of Down syndrome. It remains underdiagnosed, and delayed diagnosis remains the main factor of poor prognosis in developing countries like Madagascar. Cardiovascular exploration should be systematic in all infants with Down syndrome and those who present signs of CAV. Congenital heart disease screening centers should be increased and surgical management should be opted for at the same level for children with and without Down syndrome.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict-of-interest to be disclosed.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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