Annals of Clinical Case Reports

Case Report
Published: 04 Feb, 2022

Atrioventricular Septation Defect - A Case Report and Review of the Literature

Sofía Sierra-Poblete[1], Alejandro Bautista-Perez-Gavilan[1], Arturo Garcia-Arroyo[1], Oscar Enrique Lucio-Baez[1], Valentina Prieto-Vargas[1], Nilda Espinola-Zavaleta[2], Jorge Luis Bermudez-Gonzalez[1,2], Jose Antonio Luna-Alvarez-Amezquita[2], Arantza Marie Perez-Partida[2], Joaquin Berarducci[2], Javier Ivan Armenta-Moreno[2], Carlos Alberto Villa-Ramirez[1], Andrea Ibarra Moreno[2] and Erick Alexanderson-Rosas[1,2]*

[1]*Faculty of Medicine, National Autonomous University of Mexico, Mexico*

[2]*Nuclear Cardiology Deparment, National Institute of Cardiology Ignacio Chavez, Mexico*

Abstract

Early diagnosis and treatment of complete atrioventricular septal defects are of vital importance to avoid permanent cardiovascular disease and, more importantly, the death of a patient. This article is a documentation of the diagnosis and treatment of a patient with a common atrioventricular septation defect, referred to our institution.

Introduction

Atrioventricular septation defects are a group of congenital heart diseases that consist of a defect in endocardial cushions, also known as atrioventricular septum defect. It is characterized by the presence of a common atrioventricular union due to the absence of the membranous and muscular septum, anomalous left ventricular outflow tract, an abnormal lateral rotation of the posteromedial papillary muscle and an abnormal configuration of the atrioventricular valves [1]. This condition represents 4% to 5% of congenital heart diseases. It is more frequent in patients with Down's syndrome (40% to 45%) [2,3] and in women [4].

AV septation defect can be classified as complete or partial, and as balanced or unbalanced; it can also be catalogued with the Rastelli classification in types A, B or C [1,5-7].

Associated complications are congestive heart failure, atrial arrhythmias, complete atrioventricular block and pulmonary hypertension. The development of these complications depends on the specific morphology and the time-of-diagnosis, as well as the follow-up and treatment the patient receives [1].

Case Presentation

A 2-year-old male patient was referred to our institution with the diagnosis of systolic murmur, interventricular and interatrial communication. Previous to the admission, the patient was receiving digoxin.

The mother referred that her son had been showing fatigue during breast-feeding and profuse sweating since 6 months ago. During the previous three months, he presented two episodes of dyspnea and acrocyanosis, productive cough, fever and perioral cyanosis. He had a history of four hospital admissions due to heart failure.

Physical examination revealed distal and perioral cyanosis. In the thorax, right ventricular impulse was detected and the apex was palpated 1 cm left from the midclavicular line. Auscultation was remarkable due to a holosystolic murmur II/IV with eccentric radiation and an increased-in-intensity S2; crackles were also heard at pulmonary examination.

The Electrocardiogram (EKG) showed a left-deviated axis and a PR of 0.18 sec, biatrial enlargement, Left Ventricle (LV) enlargement and right ventricle hypertrophy (Figure 1).

Chest radiography revealed a second-grade cardiomegaly (cardiothoracic index of 0.56). Transthoracic echocardiography showed an atrioventricular septation defect. Cardiac catheterization through the left saphenous vein corroborated the presence of an atrioventricular septation defect, ostium primum interatrial communication and interventricular communication.

OPEN ACCESS

Correspondence:
Nilda Espinola-Zavaleta, Department of Nuclear Cardiology, National Autonomous University of Mexico, Juan Badiano 1, Sección XVI, Tlalpan 14080, Mexico City, Mexico,
E-mail: niesza2001@hotmail.com

Received Date: *23 Dec 2021*
Accepted Date: *11 Jan 2022*
Published Date: *04 Feb 2022*

Citation:
Sierra-Poblete S, Bautista-Perez-Gavilan A, Garcia-Arroyo A, Lucio-Baez OE, Prieto-Vargas V, Zavaleta NE, et al. Atrioventricular Septation Defect - A Case Report and Review of the Literature. Ann Clin Case Rep. 2022; 7: 2115.

ISSN: 2474-1655

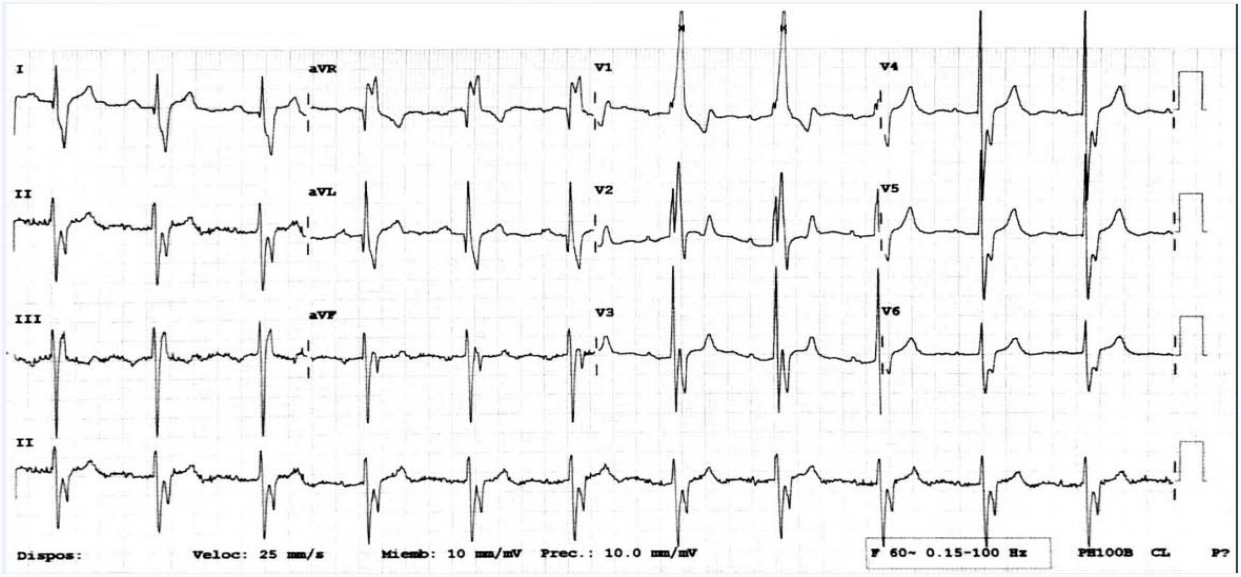

Figure 1: Electrocardiogram. Sinus rhythm with a heart rate of 72 beats per minute (bpm). Complete right bundle branch block.

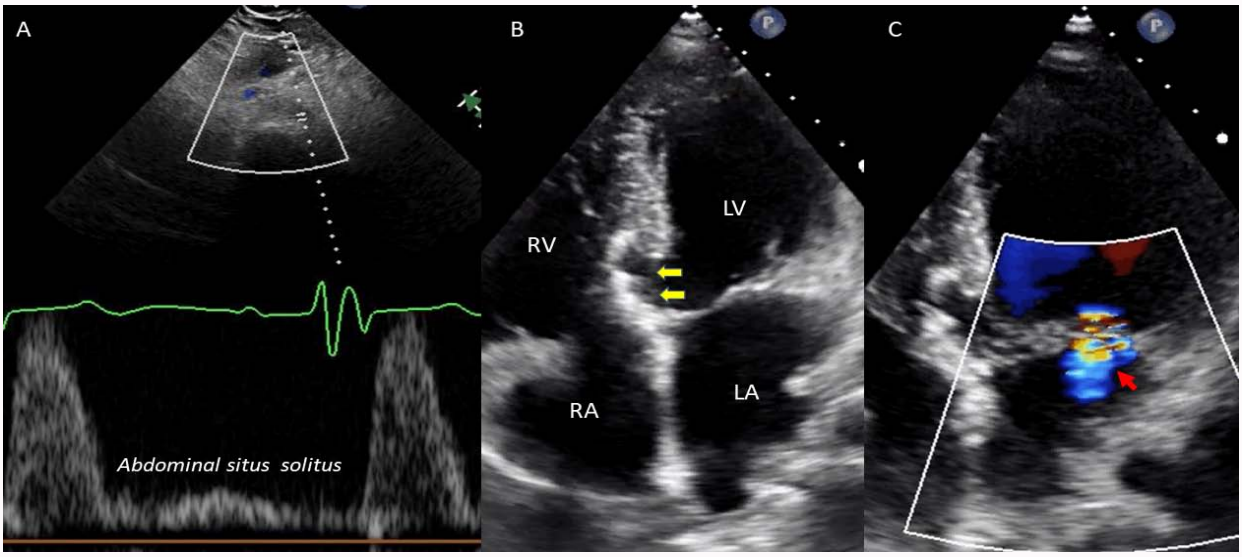

Figure 2: Transthoracic echocardiography after total correction of defect. A- abdominal situs solitus. B- Apical view of the 4 chambers with total correction of the atrioventricular septation defect (yellow arrows). C- Doppler apical view of the 4 chambers that shows mild mitral regurgitation (red arrow).
RA: Right Atrium, RV: Right Ventricle, LA: Left Atrium, LV: Left Ventricle

The patient was referred to pulmonary artery banding and through left posterior thoracotomy, the connection between the aorta and the pulmonary artery was dissected and an umbilical tape was placed.

The patient did not present any complications from the procedure. However, 4 years later, the patient was readmitted with the diagnosis of non-functional pulmonary artery banding, pulmonary hypertension, right and left atrioventricular regurgitation and complete atrioventricular septation defect Rastelli type C.

Catheterization demonstrated reversible pulmonary hypertension and the patient were considered for surgical correction. The patient continued 4 years with pharmacological management and close monitoring until total correction of the atrioventricular septation was made, total excision of the pulmonary artery banding was also realized, without complications (Figure 2). During the hospital stay, the patient received inotropic support with amrinone and dopamine.

In a follow up ECG, a first-degree atrioventricular block was detected as well as right atrial and ventricular enlargement and complete right bundle branch block. Physical examination revealed a mesosystolic murmur I/IV in the pulmonary area (which suggested pulmonary dilation) and a regurgitant murmur II/IV at the mitral area with radiation to axilla (suggesting mitral regurgitation). The patient is currently being closely followed up in our institution and shows no apparent complications.

Overview of the atrioventricular septation defects

Pathophysiology: The patient we exposed previously, had a complete atrioventricular septation defect, it combines ostium primum-type atrial communication, interventricular communication

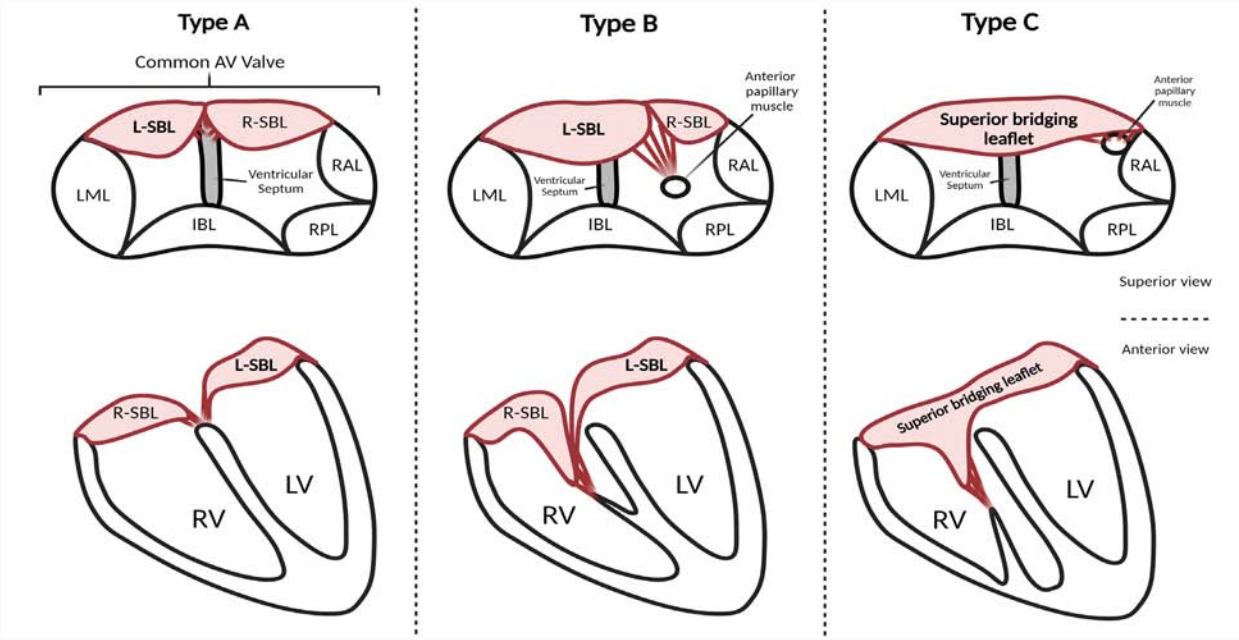

Figure 3: ABC Rastelli types with common atrioventricular valve.
RV: Right ventricle, LV: Left ventricle, R-SBL: Superior bridging leaflet (Right component), L-SBL: Superior bridging leaflet (Left component), RAL: Right Anterior Leaflet, LML: Left Mural Leaflet, IBL: Inferior Binding Leaflet, RPL: Right Posterior Leaflet. Created with BioRender.com

and a unique common atrioventricular valve [5]. A right-to-left shunt predominates due to the septation defects. Additionally, volume overload is present because of the common atrioventricular valve regurgitation, which usually is abnormal and incompetent [5,8].

Volume overload in right chambers causes increased pulmonary flow, which can lead to pulmonary hypertension and heart failure [8]. Pathological alterations at the pulmonary circulation increase the pulmonary vascular resistance and when the latter the systemic resistances, the shunt is inverted from right to left, which produces the Eisenmenger syndrome [5].

Classification: This disease is classified according to its complexity in partial or complete:

1. Partial: consists in ostium primum interatrial communication and mitral valve cleft (a space between the superior and inferior valves, above the interventricular septum).

2. Complete: consists of ostium primum interatrial communication, interventricular communication and mitral cleft.

It is important to determine whether the defect is balanced or unbalanced, according to the ventricle's dimensions. In balanced defects, the ventricles sizes are similar while in the unbalanced there is a remarkable size difference between ventricles and it must be determined which ventricle is the largest or dominant [1,5,6]

Another classification depends on the anterosuperior valves insertions according to the right ventricle, called the Rastelli classification (Figure 3) [7], which is constituted by 3 types: A, where the tendinous chordae are inserted in the ventricular septal crest; B, where the tendinous chordae are inserted in a papillary muscle (located in the union of the ventricular septum with the free wall of the right ventricle and C, where the tendinous chordae are inserted in the papillary muscle in the free wall of the right ventricle [7].

Clinical manifestations: Clinical manifestations depend on the presence and size of the interatrial and interventricular communication, as well as the competence of the atrioventricular valve [1]. The most frequent manifestation is heart failure due to right ventricle volume overload, which in the first months of life becomes apparent with the presence of polypnea, dyspnea, diaphoresis, fatigue during breast-feeding, malnutrition and failure to thrive [5,8].

Common findings on physical examination are: A simple S1 (from the common atrioventricular valve), an increased S2 (due to pulmonary hypertension), a systolic murmur due to atrioventricular valve regurgitation and a mild expulsive, systolic murmur due to pulmonary hyperflux [1,5,8]. During early stages, the presence of pulmonary hypertension and a mild pressure gradient between left and right chambers produces scarce turbulence at the shunt and thus, few findings on auscultation [5].

In cases where patients develop pulmonary vascular disease, the intensity of the expulsive murmur decreases due to the decrement in the blood flow through the pulmonary valve. However, the regurgitant murmur persists because of the atrioventricular valve [8].

If patients are over one year old, the pulmonary vascular disease produces Eisenmenger's syndrome, where the right-to-left shunt displays with progressive central cyanosis, dyspnea, syncope, desaturation and digital clubbing [5,8].

Diagnosis: The most important diagnostic test in a patient with suspicion of atrioventricular septation defect is the Doppler echocardiography, which allows to visualize directly the anatomical defect (in the four-chamber view the atrioventricular valve can be observed), the typical atrial communication and the interventricular posterior communication, as well as the functional defect [1,5]; therefore it is the election test for the diagnosis of this disease [6]. Additionally, it allows determining the grade of competence of the atrioventricular valve; it also helps estimate the right-to-left shunt and the systolic pressure of the right ventricle. Tridimensional

echocardiography is important for surgical planning [1,5,6].

An ECG must be performed in all the patients where a septation defect is suspected. The main finding is left axis deviation [5]. Furthermore, atrial flutter could be detected and, in older patients, complete atrioventricular block associated with right ventricle enlargement and right bundle branch block could be identified [1,6]. Existence of pulmonary hypertension could manifest as a wide P wave, which can also be altered due to right ventricle overload [6].

Chest radiographs exhibit cardiomegaly with prominent right atrium, right ventricle dilation and increased pulmonary flow. In cases of mild interatrial communication and important incompetence of the atrioventricular valve, cardiomegaly may manifest due to left ventricle enlargement with normal pulmonary flow. Moreover, signs of Eisenmenger's syndrome can be detected, such as pulmonary artery calcification, dilated pulmonary arteries with narrowing of peripheral vessels [1,6].

Cardiac catheterization directly estimates the pulmonary artery pressure to determine the severity of pulmonary hypertension. In addition, it allows to evaluate the pulmonary vessels reactivity through the administration of a vasodilator agent, aiming to define if the pulmonary resistances are reversible and, consequently, if the patient is a good candidate for surgical management. Nowadays, cardiac catheterization has been almost completely replaced by transthoracic echocardiography and its only application is in patients that are in late stages of the disease, when there is an advanced pulmonary vascular alteration [1].

Treatment: A surgical procedure should be performed in patients with a de novo or non-corrected atrioventricular communication. Additionally, surgery is indicated in patients with symptomatic persistent regurgitation (or postsurgical stenosis) of the mitral valve, or patients with atrial arrhythmias, ventricular function impairment or with severe subaortic obstruction (mean rest gradient >50 mmHg) [1,6]. If possible, surgical correction should be performed before the six months old, aiming to avoid pulmonary vascular disease [5,6].

In cases of severe pulmonary hypertension, there must be a left-to-right shunt of 1.5:1 as minimum and signs of pulmonary vascular reactivity to a vasodilator agent, such as oxygen, nitric oxide or prostaglandins [1].

Our patient was first managed with a pulmonary artery banding followed by intracardiac correction, called "Stepped approach". This approach has been replaced by primary intracardiac correction during breast-feeding, pretending to achieve adequate atrial and ventricular septations, as well as to reconstruct the left and right atrioventricular valves. The employed techniques are simple-patch, double patch and no-patch to close the interatrial and interventricular communications. Occasionally, a mitral valve substitution could be needed when the correction is not feasible [1,6].

Prognosis: Surgically-managed patients need frequent evaluations due to the recovery rate at 5 years of only 75% [1]. Mortality attributed to surgery also depends on the malformation anatomical characteristics, the patient's functional status, and the severity of pulmonary hypertension [5]. After the procedure, late onset complications can occur, for example patch dehiscence, residual septal defects, atrioventricular block (which outpatient developed), atrial flutter or fibrillation, mitral valve dysfunction and subaortic stenosis [1,5,6]. As a post-operatory evaluation, a Doppler Echocardiography, an ECG, and chest radiography should be performed to estimate cardiac function after surgery [5].

Patients that are not surgically treated have a higher risk of death than those surgically treated in the first year of life, mostly due to heart failure and its complications [5]. Those who survive, develop Eisenmenger's syndrome, and usually develop congestive heart failure in the fifth decade. The most common death causes are sudden death in (30%), congestive heart failure in (35%), and hemoptysis in (15%). Perioperative deaths during extracardiac surgery can happen, as well as infectious events such as cerebral abscesses and endocarditis. Death can also occur during gestational period in some cases.

Conclusion

Patients with a complete atrioventricular septation defect should be diagnosed and managed in a prompt and comprehensive manner to correct the anatomical and functional defects that this disease exhibits. It is important to highlight the fact that, in case of not performing surgical treatment, complications can shorten the lifespan of patients as well as impair their quality of life. Multidisciplinary assessment and management of different cardiology subspecialties improve the prognosis of patients with this congenital heart disease.

References

1. Braunwald E, Zipes D, Libby P, Bonow R, Mann D, Tomaselli G. Braunwald's heart disease: A textbook of cardiovascular medicine. 11th Ed. Philadelphia, PA: ELSEVIER; 2018.
2. Fyler DC, Buckley LP, Hellenbrand WE. Endocardial cushion defect: Report of the New England Regional Infant Cardiac Program. J Pediatr. 1980;65(2):441-4.
3. Freeman S, Taft L, Dooley K, Allran K, Sherman S, Hassold T, et al. Population-based study of congenital heart defects in Down syndrome. Am J Med Genet. 1998;80(3):213-7.
4. Freeman S, Bean L, Allen E, Tinker S, Locke A, Druschel C, et al. Ethnicity, sex, and the incidence of congenital heart defects: A report from the National Down Syndrome Project. Genet Med. 2008;10(3):173-80.
5. Casaldáliga J. Endocardial cushion defects. Spanish Society of Pediatric Cardiology and Congenital Heart Disease.
6. Lince Varela R. Cardiología pediátrica. 2nd Ed. España: Editorial Médica Panamericana; 2013.
7. Kuri Nivon M, Martínez Martínez E, Muñoz Castellanos L, Espinola-Zavaleta N. Defecto septal atrioventricular. Estudio anatomopatológico y correlación embriológica. Archivos de Cardiología de México. 2008;78(1):19-29.
8. Craig F, Aykut T. Clinical manifestations, pathophysiology, and diagnosis of atrioventricular canal defects. 2018.