Continuous heart murmur: a sign of inestimable value

Dr. Hiram Tápanes Daumya, MD; Maylín Peña Fernández, MD; and Andrés Savío Benavides, MD

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Acronyms
APW: aortopulmonary window
PDA: patent ductus arteriosus

INTRODUCTION
Heart murmurs are commonly detected during routine physical examination in the pediatric population and are the most prevalent sign of cardiopedia-trics. Therefore, it is the leading cause of referral to pediatric cardiologists at Boston Children's Hospital. The blood flow through the cardiovascular system is silent because the current is laminar as blood columns. Heart murmurs are the result of turbulence
in the current flowing at high speed into or out of the heart, causing audible vibrations that reach frequencies between 20 and 20 thousand hertz\(^5,6\).

Three key factors determine the production of turbulence (murmurs), they are:

1. Increasing flow through normal or abnormal valves in anterograde direction (normal direction).
2. Forward flow through an irregular stenotic valve, or towards a dilated vessel (normal direction).
3. Backward flow through an insufficient valve or congenital defect\(^5\).

In general and considering the moment of the cardiac cycle in which murmurs appear, these can be classified into:

- **Systolic murmurs:** They are divided into mesosystolic-ejective (aortic and pulmonary foci), and regurgitant that are auscultated at the level of the mitral and tricuspid atrioventricular valves; also those related to interventricular septation defects belong to this subgroup.
- **Diastolic murmurs:** They are subdivided in regurgitant that are present in pulmonary and valvular aortic insufficiency cases, and those that relate to the rapid ventricular filling (mid-diastolic murmurs) and presystolic atrial filling.
- **Continuous murmur:** those auscultated in both phases of the cardiac cycle and whose origin is represented by a long list of multiple causes.

In this general classification of heart murmurs some authors include:

- **Systolic-diastolic murmurs:** They can also be auscultated in both phases of the cardiac cycle but the second heart sound between them is defined, while in the continuous the second noise is masked by the murmur. The origin of systolic-diastolic murmurs is derived from the overlapping of diseases such as ventricular septal defect with aortic insufficiency, pulmonary or double aortic injury, common trunk with valvular insufficiency, among others\(^7\).
- **Innocent murmurs:** They have a frequency of 60-85% in children between three and six years old, and are characterized by their short duration, systolic phase onset, low intensity, low irradiation, intensity change with the position and enhancement with hyperdynamic circulatory changes. In these cases, telecardiogram and electrocardiogram are normal. Among the most common causes are Fogel’s pulmonary murmur, aortic innocent murmur, Still’s vibratory murmur and Evans’ late murmur\(^8-11\).

Other authors include these functional or innocent murmurs in the systolic group, having in mind that indeed all these occupy the systolic phase of the cardiac cycle.

In order to clarify the pathophysiological, causal and semiological aspects of a continuous murmur this literature review is conducted, which describes the fundamental clinical elements of the different diseases that can cause continuous murmurs, as well as the criteria for clinical, hemodynamic or surgical approaches.

**CAUSES FOR CONTINUOUS MURMUR**

Continuous murmurs are those that persist and are heard in systole and diastole, and are caused by the continuous flow of blood from an area of high pressure to one of low pressure, maintaining a pressure gradient throughout the cardiac cycle\(^8\).

An essential element in the pathophysiology of continuous murmur is the hemodynamic phenomenon responsible for maintaining a pressure gradient between the vessels, and capable of maintaining a turbulent flow which covers the systole and a segment of cardiac diastole, and masks the second cardiac sound.

Some authors classify continuous murmurs according to the structures involved in the shunt and divide them into: arterio-arterial, arterio-venous and veno-venous\(^12\).

The classical semiological description of a continuous murmur was made by Gibson in 1900 in relation to the murmur present in patent ductus arteriosus (PDA), referring to this as a machinery murmur. However, the diseases that make the causal list of the continuous murmur are multiple and all of them are especially relevant for the pediatric cardiologist.

In general, the causes of continuous murmur are:

1. PDA.
2. Aortopulmonary window (APW).
3. Ruptured aneurysm of sinus of Valsalva.
4. Classic or modified Blalock - Taussig shunt.
5. Coronary arteriovenous fistulas.
6. Truncus arteriosus.
7. Venous hum.
8. Mammary souffle.

All causes of continuous murmur except for the mammary souffle, appear in childhood, and are associated with congenital malformations or palliative procedures related to these.

**Patent ductus arteriosus**

As the name suggests it is caused by postnatal persistence of the ductus arteriosus, which is an embryological structure originating from the distal left sixth aortic arch and connects the pulmonary artery trunk, near the left pulmonary artery branch, with the descending thoracic aorta of the left subclavian artery after birth. The existence of the ductus arteriosus is essential for intrauterine life, as 60% of the cardiac output passes through it.

The first accurate description of the ductus arteriosus was made by James Gibson in 1900, however, William Harvey had already made excellent anatomical descriptions in the sixteenth century. For its size and hemodynamic effects PDA is classified as:

- Large: when it measures over six millimeters at both ends (aortic and pulmonary).
- Moderate: when it measures three to six millimeters at both ends (aortic and pulmonary).
- Small: when measures less than three millimeters at both ends.

According to the angiographic morphology of the duct, Krishenko (1989) classified them into:

- Type A: cylindrical, flattened, and semi-closed towards the pulmonary end.
- Type B: cylindrical, flattened, and semi-closed towards the aortic end.
- Type C: cylindrical, straight without recesses.
- Type D: cylindrical, with lung and aortic double recess.
- Type E: Long with recess close to the lung one.

Its clinic is booming from the third month of life, when pulmonary pressures fall, and is mainly characterized by tachypnea, repeated broncho-catarrhal episodes, celer-type bulging arterial pulses, continuous murmur in I-II left intercostal space, rumbling and left gallop rhythm.

Follow up of continuous murmur in this disease is extremely important for the pediatric cardiologist since its absence, or the only presence of the diastolic component, together with a pulmonary component of the second hyperphonetic sound should lead to suspect the presence of pulmonary hypertension, which in extreme circumstances leads to the Eisenmenger syndrome, that determines the impossibility of the definitive defect correction.

Diagnosis is complemented by the electrocardiogram that can vary over a spectrum that goes from normal in small PDA to biventricular growth in that of large hemodynamics impact. When Eisenmenger syndrome develops some signs of right ventricular hypertrophy appear with QRS axis to the right, tall R in V1 and V2 and right Sokoloff-Lyon signs (R of V1-V2 + S of V5-V6 greater than 11 millimeters).

In telecardiogram some signs of left ventricular hypertrophy with a domed pulmonary artery trunk and increased pulmonary flow are observed, although in small PDA the telecardiogram can approximate the normal. In Eisenmenger syndrome pulmonary oligohemia is associated with right ventricular growth. Echocardiography confirms the diagnosis.

Medical treatment is focused on the control of heart failure with diuretics, aldosterone inhibitors, digoxin, angiotensin converting enzyme inhibitors or angiotensin II–receptor inhibitors.

Its definitive treatment, depending on the protocols that the different institutions use, may be catheterization (percutaneous coronary intervention) or surgery, which is usually simple and free of morbidity and mortality, with rare exceptions.

**Aortopulmonary window**

It is a direct communication between the ascending aorta and the pulmonary artery trunk, resulting from a partial failure in the development of aortic-pulmonary septal complex, a structure that in its embryological origin divides the great vessels.

One might state that APW is a defect that occupies an intermediate position between normality and truncus arteriosus (total deficit in the formation of aortic-pulmonary septal complex), from which it differs since in APW there are two semilunar valves: the aortic and pulmonary valves, whereas in the truncus arteriosus there is only one sigmoid or trunk valve and a single vessel leaving the heart, from which, depending on the
anatomical variant, the pulmonary branches and supra-aortic trunks are born in a different position.

This congenital heart disease was first described in 1830 by Elliotson in an anatomical specimen, while Cooley performed the first successful corrective surgery in 1957. APW is a really rare malformation with an estimated incidence of 0.2 to 0.6% of all congenital heart diseases. There are various classifications and the most accepted is that of Mori et al, which divide them into:

- Type I or proximal: the defect is circular in an equidistant area between the sigmoid valve and the bifurcation of the pulmonary branches. It constitutes 70% of cases.
- Type II or distal: It has spiral shape and affects the trunk and the origin of the right pulmonary artery. It constitutes 25% of cases.
- Type III: Complete aortopulmonary septum defect. It represents the remaining 5%.

Its symptoms appear around the first month of life and are characterized by dyspnea (occurring with feedings), sweating, growth retardation and respiratory distress.

On physical examination tachycardia and tachypnea are predominant. The signs found on auscultation are very similar to those of PDA with continuous murmur, mitral rumble and left gallop, but unlike PDA there are early signs of irreversible pulmonary hypertension. This situation requires surgical correction before 6 months of age, although there have been closures, in specific circumstances, by interventional catheterization.

Ruptured aneurysm of sinus of Valsalva

The sinuses of Valsalva are aortic wall dilations located between the aortic annulus and the sinotubular junction. Its location is related to the coronary arteries; therefore they are designated as right or left coronary sinus, and non-coronary sinus. Aneurysm of Sinus of Valsalva is defined as the dilation of one of them with loss of continuity between the middle layer of the aortic wall and the valve annulus.

The incidence of this heart disease is estimated between 0.15 and 1.5%. Some authors suggest that there is an ethnic variability in the presentation of this type of disease; hence it is known that it is five times more common in Asians than in Western countries, and is presented with a male to female ratio of 4:1.

The cause of the aneurysm can be congenital or acquired, in turn, acquired forms can be traumatic, infectious (primarily syphilitic aortitis) or due to degenerative diseases. Right sinus aneurysmal rupture is much more frequent (65 to 86% of cases), followed by the non-coronary (between 10 and 30%) and finally, left sinus rupture with 2 and 5% cases.

Aneurysm rupture towards the cardiac chambers occurs approximately in the following order of frequency, right ventricle (60%), right atrium (29%), left atrium (6%) and left ventricle (4%).

Extracardiac ruptures in the pericardium or pleura are extremely rare, but fatal, usually due to aneurysms of acquired origin.

The pathophysiology of this condition depends primarily on the volume circulating through the communication, on the rapidity with which it sets, on the cardiac chamber with which the exchange is established and the functionality of such chamber. Therefore, a variety of symptoms and signs can be present: cough, fatigue, chest pain, dyspnea, arrhythmias, and coronary artery compression with signs of ischemia, with an always present continuous murmur. However, when perforation is gradual this can acceptably be tolerated in 25% of cases.

The only formal classification for ruptured aneurysm of sinus of Valsalva was made by Sakakibara and Konno in 1962 (Table 1). Taking into account the affected coronary sinus and the area to which it bulges or breaks, four types are identified.

<table>
<thead>
<tr>
<th>Type</th>
<th>Connection</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Right SV with the outflow tract of the right ventricle below the pulmonary valve</td>
</tr>
<tr>
<td>II</td>
<td>Right SV with right ventricle in the supraventricular crest</td>
</tr>
<tr>
<td>IIIa</td>
<td>Right SV with the right atrium</td>
</tr>
<tr>
<td>IIIv</td>
<td>Posterior area of right SV with the right ventricle</td>
</tr>
<tr>
<td>IIIa+v</td>
<td>Right SV with atrium and the right ventricle</td>
</tr>
<tr>
<td>IV</td>
<td>Non-coronary SV with right atrium</td>
</tr>
</tbody>
</table>

SV= Sinus of Valsalva

Tabla 1. Clasificación de Sakakibara y Konno de los aneurismas del seno de Valsalva.
About 20% of congenital aneurysms of the sinus of Valsalva are not perforated and they are discovered at autopsy or at surgery for a coexisting ventricular septal defect. When a ruptured or intact aneurysm of Valsalva penetrates the interventricular septum base it can cause a complete heart block which can cause death or syncope.  

This disorder is often linked to other congenital defects, primarily with interventricular communication, most often in type I or supracrestal it may also be associated with aortic regurgitation (41.9%), pulmonary stenosis (9.7%) or aortic (6.5%), aortic coarctation (6.5%), PDA (3.2%) and tricuspid regurgitation (3.2%).

Most patients without rupture remain asymptomatic. When rupture occurs, usually between the second and third decades of life, the clinical presentation is usually acute chest pain or heart failure data. If the shunt magnitude is not significant, these patients can be stabilized for a few days, however, they develop progressive aortic insufficiency and their survival without surgery is limited, with an average of 3.9 years.

The electrocardiogram may vary between normality and the presence of QRS axis to the right and tall R in V1, with the corresponding ST and T depression due to right ventricular volume overload; signs of myocardial ischemia are much rarer. The telecardiogram may show a mild cardiomegaly by hypertrrophy and dilation of the right cavities and increased pulmonary flow, with a bulged pulmonary artery trunk. All this will depend on the degree of hemo-dynamic impact of the heart disease.

Echocardiography has a diagnostic accuracy between 75 and 90% in both ruptured and intact aneurysms. With this method we can discriminate the size, sinus of origin, termination or drainage site, severity, valvular regurgitation mechanism, cardiac or vascular abnormalities associated, which are very important data for surgery. The angiographic study, considered the baseline examination, is rarely necessary for diagnosis. CT scan and MRI offer high sensitivity and specificity, but their use for the diagnosis of this type of condition is very limited.

Due to disease evolution all patients should be surgically treated. The first intervention of this kind took place in 1955, under deep hypothermia without cardiopulmonary bypass. Surgical treatment of aneurysm of the sinus of Valsalva is safe, with a mortality of about 1%. Some authors suggest that perioperative mortality increases 4-5 times in cases of infection or endocarditis. Au et al. conducted a review of 53 patients operated on between 1978 and 1996, and reported satisfactory results with large long-term survival, absence of early perioperative deaths and recurrences after initial repair.

**Blalock-Taussig Surgical Shunt**

This procedure is named after its creators, two American doctors, Helen Brooke Taussig, for many "Mother of cardiopediaitrics" and the surgeon Alfred Blalock, who performed an extraordinary act on November 29, 1944, when created a palliative communication between the subclavian artery and a pulmonary artery branch. The joy of being the first to benefit from the procedure was for Eileen Saxon, and this milestone was multiplied by thousands and gave hope of life to countless patients. According to Jaramillo, Potts and colleagues in 1946 described an anastomotic procedure between the descending aorta and left pulmonary artery branch, while Waterston in 1962, published a technique that consisted of anastomosing the ascending aorta to the right pulmonary artery and other techniques that emerged in time. The techniques of Potts and Waterston have practically fallen into disuse for causing excessive pulmonary flow, left heart failure, obstructive pulmonary hypertension, and distortion problems when making definitive intracardiac repair and due to an increase in mortality of up 40% in relation to the Blalock-Taussig shunt.

This classic procedure was modified in 1980 according to a proposal made by McKay and colleagues, and a synthetic material such as the polytetrafluoroethylene type began to be used; grafts from 3-5 mm were used and a number of advantages with the modified procedure were recognized:

- High early patency.
- Shunt regulation by systemic artery size.
- Preservation of the subclavian artery for future definitive correction.
- Relative ease of surgical procedure in relation to the classic one.
- Easiness to interrupt the shunt when the complete repair is done.

The disadvantages of the modified Blalock-Taussig procedure are:

- Polytetrafluoroethylene is not an optimal material,
prone to the formation of “pseudointima” and late obstruction, especially when they are from 3 to 3.5 mm.

Distortion of the pulmonary artery anatomy by use of rigid and thick shunted material to small or thin branches.

Despite created more than 65 years ago, the Blalock-Taussig shunt, with some modifications now, remains a palliative procedure, essential for patients with cyanotic congenital heart disease and pulmonary flow alteration and therefore of alveolar pulmonary hematosis.

**Truncus arteriosus**

The truncus arteriosus is a congenital heart disease characterized by a single arterial trunk coming from the heart, and gives rise to the coronary arteries, the pulmonary arteries (or at least one) and brachiocephalic arteries. The unique truncus arteriosus usually originates from two distinct ventricles and is accompanied by a large ventricular septal defect with a single trunk and malformed valve. It is a rare heart disease. The prevalence is 0.03 per 1,000 live births and occupies 0.7 to 1.4% of congenital heart defects. In the embryology of this malformation a disorder in the septal aortic-pulmonary complex formation occurs, so the truncus arteriosus is maintained as a single structure from which the coronary arteries emerge, the pulmonary arteries and supraaortic trunks, so a mixture of arterial and venous blood is produced with consequent cyanosis and increased pulmonary flow, which can lead to pulmonary hypertension.

According to Somoza, the first description of this heart disease is attributed to Wilson in 1798, and the important contributions of Buchanan and Preisz are highlighted.

In the early twentieth century multiple classifications of this heart condition appeared, and those of Collet and Edwards (1949) and Van Praagh (1965) are the most commonly used (Table 2).

The typical clinical presentation is that of congestive heart failure that starts in the first weeks of life. Parents report that the child tires easily with feedings, and presents tachypnea and profuse diaphoresis, there is usually evident cyanosis at birth and this increases with age. On physical examination, we can have a first normal sound with loud opening crack and the second sound is unique. There may be high frequency diastolic pulmonary murmur due to trunk valve insufficiency. The continuous murmur in this condition is rare, due to the appearance of aorto-pulmonary collaterals or when ostial stenosis of the pulmonary branches is present.

The electrocardiogram and X-ray show signs of right or biventricular growth, with increased pulmonary flow and ovoid morphology. The echocardiogram shows the emergence of the trunk vessel overriding the interventricular septum, which serves as a roof, also both pulmonary arteries originating from it can be seen, and the location of the aortic arch (right or left) can be determined.

The presence of this disease is a strict and absolute indication for surgery, which is usually performed at around 3 months of age, for as long as the patient’s condition allows, pulmonary artery pressures are ex-

<table>
<thead>
<tr>
<th>Collet and Edwards Type</th>
<th>Description</th>
<th>Van Praagh Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>There is a common pulmonary trunk from the trunkal artery</td>
<td>A1</td>
<td>It matches type I of Collet and Edwards</td>
</tr>
<tr>
<td>II</td>
<td>Independent origin, but very close to the pulmonary arteries</td>
<td>A2</td>
<td>Includes types II and III of Collet and Edwards</td>
</tr>
<tr>
<td>IIIa</td>
<td>Independent and distant origin of both pulmonary arteries</td>
<td>A3</td>
<td>Absence of pulmonary branch (hemitrunk)</td>
</tr>
<tr>
<td>IIIv</td>
<td>Pulmonary branches emerging from the descending aorta (for some it is considered an extreme form of Fallot with pulmonary atresia)</td>
<td>A4</td>
<td>Truncus associated with interrupted aortic arch</td>
</tr>
</tbody>
</table>
pected to drop. To ensure continuity of the pulmonary vasculature and the right ventricle, valve conduit with certain characteristics are used whose prognosis and durability depend primarily on: a) performing surgery at an earlier age, even during the first month of life, b) improvement of cardiopulmonary bypass techniques and myocardial protection in the neonate c) improvement in postoperative care of patients. For simple forms postoperative mortality is 5 to 10%, while in the complex forms it can reach 20%\textsuperscript{51}.

**Coronary arteriovenous fistulas**

In the context of congenital anomalies of coronary arteries, fistulas are classified as termination anomalies and constitute between 0.2% and 0.4% of congenital heart disease\textsuperscript{52}, despite its low incidence it is the most frequent of coronary anomalies, so it can be found in 0.15% of coronary angiograms performed\textsuperscript{53}. It is one of the most common congenital malformations of coronary circulation allowing survival into adulthood\textsuperscript{54,55}.

The first description of these fistulas was published by Krause in 1865 and mentioned by Abbot\textsuperscript{57} in 1906, and the first surgical correction was performed by Björk y Crafoord\textsuperscript{58} in 1947.

Regarding the fistulous structure site of origin, the right coronary artery constitutes 55% of cases, left coronary artery 35% and 5% for both. 90% of fistulas end on the right side of the heart, in order of frequency: in the right ventricle, right atrium, the coronary sinus and the pulmonary circulation, they rarely end in the left ventricle\textsuperscript{52,59}.

Patients with this disease may remain asymptomatic until adulthood. When symptoms occur, the most common are: angina pectoris by coronary stealing, dyspnea by pulmonary hypertension, manifestations of infective endocarditis or heart failure\textsuperscript{59,60}. Anecdotally, hemoptysis has been found\textsuperscript{61}.

The most important findings on physical examination (when present) are the continuous murmur and signs of heart failure, pulmonary hypertension or myocardial ischemia.

Diagnosis is mostly supplemented by two-dimensional color Doppler echocardiography which gives us the essential data of the anatomy and pathophysiology of the lesion\textsuperscript{62}. Cardiac catheterization is the study of choice to define the anatomy of the coronary anomaly and its hemodynamic effect and to define existing cardiac abnormalities or the presence of coronary obstruction. Other echocardiographic modalities are also useful, for example contrast CT scans and MRI\textsuperscript{62,63}.

Regarding final management opinions are divided, some authors recommend the closure of all fistulas in childhood, even if asymptomatic, others, however, propose that only symptomatic patients should be treated or those at risk of complications\textsuperscript{64,65}. The percutaneous treatment is proposed today as the elective method, less radical and a shorter hospital stay\textsuperscript{66}, and surgery is reserved for cases with multiple fistulas when the fistulous path is narrow and restrictive and drains into a cardiac chamber, or when complications arise during coils embolization\textsuperscript{67-69}.

**Venous Hum**

According to Zarco\textsuperscript{5}, it was described by Potain in 1867 as a continuous innocent or benign murmur of low-frequency sound that is auscultated in children, mainly between 3 and 6 years old. It is due to the passage of blood at high speed through the veins of the neck, and is auscultated more often in the right supraclavicular fossa, less frequent in the left, in a sitting position; with light pressure on the neck its features are enhanced, and disappears with head movements, or decubitus position with strong pressure of jugular veins. Interestingly it is not enhanced around the second sound, but its peak is mid-diastolic\textsuperscript{5,7}, its tone is rough and noisy, and it is believed to be due to the deformation caused by the transverse cervical apophysis in the internal jugular vein which can affect the laminar flow at this level\textsuperscript{12}.

**Mammary souffle**

During pregnancy there are a series of physiological changes characterized by increased plasma volume, increased heart rate and decreased peripheral resistance, starting from the sixth week of fetal life, reaching a peak between 20 and 24 weeks and are maintained until the end of pregnancy\textsuperscript{70,71}. As part of the resulting hyperdynamic state various types and locations of heart murmurs can be auscultated, and due to the increased blood flow in a breast preparing for future breastfeeding, a mammary souffle or continuous mammary murmur\textsuperscript{70} is auscultated, which as semiological characteristics presents a fading with the stethoscope’s pressure, it also disappears when the
pregnant woman stands up, and its maximum intensity towards the end of pregnancy, and may even remain in the postpartum period.\footnote{71}

**CONCLUSIONS**

The continuous murmur is a sign with its own characteristics, with a well-defined origin and pathophysiology, that complemented with appropriate physical examination, electrocardiogram and telecardiogram can guide us to the diagnosis of some diseases that characterize it. Its recognition and proper interpretation are key elements in our daily work.

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