Intermittent Disappearance of the Murmur of Patent Ductus Arteriosus


The murmur of patent ductus arteriosus is usually considered to be stable, although it may be modified by certain pharmacologic agents. Absence of the typical ductus murmur has most commonly been ascribed to the presence of relative or absolute pulmonary hypertension, occasionally it is ascribed to heart failure, a very small ductus, or spontaneous closure of the ductus. Spontaneous disappearance and subsequent reappearance of the typical murmur of patent ductus unassociated with pulmonary hypertension, heart failure, polycythemia or other congenital anomalies has not been previously documented to our knowledge. The purpose of this report is to describe this phenomenon and to attempt to explain it.

Case Report

The patient, a 9-year-old girl, was studied because of a heart murmur. She was born after 7 months’ gestation, weighing about 1,700 Gm. A murmur was discovered at age 2 but, according to the mother, it was not always heard. The child had frequent upper respiratory tract infections and was said to tire more easily than other children. There was no history of cyanosis, edema, squatting, fainting, or significant retardation in growth and development.

It became clear that the patient could tell whether or not the thrill accompanying her murmur was present. She stated that her “washing machine” would disappear with quiet rest and return when she became excited.

The patient’s immediate family, including 3 younger sisters, were in good health. A younger brother had died within a few days of birth of congenital anomalies of the spinal cord. A maternal aunt was thought to have had some form of congenital heart disease and had died at age 42.

The patient weighed 54 pounds and was 49 inches tall (both in the fifteenth percentile). She was alert, well developed, slender, and slightly pale. Clubbing, edema, cyanosis, and neck-vein distention were not present. The lungs were clear. The point of maximum impulse was at the midclavicular line in the fifth intercostal space. The second sound in the pulmonic area was palpable, and at times a long thrill could be felt in the pulmonic area. The pulmonic second sound was louder than the aortic second sound. In the pulmonic area and for some distance about it could be heard a continuous murmur with a rough systolic component of maximum intensity at the time of the second sound and continuing through diastole as a softer, higher pitched murmur. The peripheral pulses were bounding and cuff blood pressure in the left arm was 107/52.

During several examinations the murmur abruptly disappeared for varying lengths of time (fig. 1). This usually occurred during quiet rest or sleep. When the bruit was inaudible, cuff blood pressures averaged 101/75. After periods of time, as long as 30 minutes, the murmur and thrill returned.

Tests of blood and urine and an electrocardiogram were normal. X-rays of the chest demonstrated minimal prominence of the pulmonary artery segment, but otherwise the cardiac contour and lung fields were unremarkable. At fluoroscopy the heart appeared hyperdynamic. Along the right border of the heart there appeared to be intrinsic expansile pulsations of the pulmonary vessels. The left ventricle did not completely clear the spine on deep inspiration in the left anterior oblique position.

The patient was studied on many occasions. Right heart catheterization was carried out under intravenous Pentothal anesthesia. Pressures were obtained with an electromanometer and recorded on a multichannel direct-writing apparatus. Blood oxygen determinations were made according to the method of Van Slyke and Neill. Duplicate determinations agreed to within 0.2 to 0.3 volumes per cent. Phonocardiograms were recorded in the log position on a Sanborn Twin-Beam Cardielle. Determinations of arterial PCO₂, expired PCO₂,

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and tidal volume were performed in order to calculate physiologic dead space from the Bohr equation.

At thoracotomy a patent ductus arteriosus was found, divided, and sutured without difficulty. The patient recovered without incident and 2 months later she was reported to be less easily fatigued and eating unusually well.

**Observations**

**Cardiac Catheterization**

Data obtained at right heart catheterization were consistent with the clinical diagnosis of patent ductus arteriosus (table 1). Before the procedure was begun, the murmur was inaudible. The murmur returned as the procedure started and remained throughout the study. A significant oxygen rise occurred at the pulmonary artery level and the pulmonary blood flow was more than twice systemic blood flow. Pressures in the pulmonary artery and right ventricle, as well as the calculated pulmonary vascular resistance, were well within the range of normal. Brachial artery pressure was normal.

![Figure 1](http://circ.ahajournals.org/)

**Figure 1**

Top, Typical continuous murmur usually heard in this patient. Bottom, Recording in same area at same sensitivity demonstrating spontaneous disappearance of the murmur.

**Table 1**

<table>
<thead>
<tr>
<th>Location</th>
<th>S.D.</th>
<th>Mean</th>
<th>Oxygen content per cent</th>
<th>Per cent oxygen saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>5/0</td>
<td>3</td>
<td>10.2</td>
<td>73</td>
</tr>
<tr>
<td>Right atrium</td>
<td>6/3</td>
<td>4</td>
<td>9.7</td>
<td>69</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>21/2</td>
<td>6</td>
<td>9.6</td>
<td>69</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>21/13</td>
<td>16</td>
<td>12.9</td>
<td>92</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>20/13</td>
<td>16</td>
<td>12.0</td>
<td>86</td>
</tr>
<tr>
<td>Pulmonary <em>capillary</em></td>
<td>15/5</td>
<td>10</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Left brachial artery</td>
<td>118/73</td>
<td>90</td>
<td>13.8</td>
<td>99</td>
</tr>
</tbody>
</table>

Systemic flow (O₂ consumption assumed to be 160 ml/min./M²=3.5 L/min.

Pulmonary flow (with use of RPA O₂ content)= 7.9 L/min.

Pulmonary vascular resistance=138 dynes/sec/em.²

*Oxygen capacity of blood=14.0 volumes per cent.

**Phonocardiographic Studies**

Repeated attempts were made to demonstrate a particular posture or other state that was consistently associated with disappearance of the murmur. On one occasion it was found to be absent while the patient was standing, but generally it disappeared with the patient quietly resting or sleeping in the supine position. Assumption of the latter attitude, however, was not invariably followed by disappearance of the bruit. Knee-chest, prone, Trendelenburg, Fowler, 60-degree upright tilt, and various lateral positions did not affect the murmur.

The murmur was observed to disappear either abruptly or gradually. When the disappearance was gradual, the diastolic portion would shorten and disappear, then the systolic component would progressively diminish in intensity until it became inaudible. For periods varying up to 30 minutes normal heart tones only would be heard. Reappearance was also gradual or abrupt (fig. 2). Marked respiratory variations in the murmur were sometimes noted upon its return, i.e.,
a loud continuous murmur in expiration, near absence to absence during inspiration followed by re-establishment and no noticeable changes throughout the respiratory cycle. A thrill was present when the murmur was most pronounced; when the murmur was soft or absent, no thrill was palpable.

The effects of various agents were tested. No agent produced complete disappearance like what occurred spontaneously. Anxiety (increased cardiac output, heart rate, and stroke volume)\textsuperscript{12} induced by impeding and actual needle pricks (to administer local anesthesia for arterial puncture) made the murmur louder. Inhalation of 100 per cent oxygen (decreased pulmonary vascular pressure)\textsuperscript{2, 13, 14} and carotid sinus massage produced no consistent change in the murmur. Inhalation of 13 per cent oxygen for 5 minutes (increased pulmonary vascular pressure)\textsuperscript{2} brought out marked respiratory variations in the bruit. These were manifest as striking diminutions in amplitude during inspiration. Amyl nitrite inhalations (systemic vasodilatation)\textsuperscript{15} produced a decrease in amplitude and shortening of the diastolic portion of the murmur at the onset of the facial flush on one occasion. On a second occasion amyl nitrite transiently produced the same effect at the height of the flush.

\textbf{Figure 2}

Top. Inaudible, low-pitched vibrations are present in systole. The murmur partially returns during one cycle. Bottom. Abrupt return of the murmur; it disappeared again after several cycles.
That the murmur itself might have been an unusually loud venous hum was ruled out by the complete lack of effect of movements of the head and unilateral or bilateral jugular venous compression.

Arm blood pressures were repeatedly determined with a pediatric sphygmomanometer. The mean of 8 determinations on several occasions when the murmur was absent was 101/75 (range 98-105/64-80, mean pulse pressure of 26). The mean of 5 determinations when the murmur was present was 107/52 (range 100-110/44-65, mean pulse pressure of 55). Despite its limitations as compared with direct intra-arterial pressure recordings, repeated external cuff pressures have value since they were obtained by a single observer. The narrowed pulse pressure, largely due to an increase in diastolic pressure, observed when the murmur was absent, suggested that the shunt through the patent ductus arteriosus might have diminished greatly at these times.

Physiologic Dead Space Studies

In 1959, Riley et al.16 reported that assumption of the upright position in normal subjects resulted in an increase in physiologic dead space due to lack of perfusion of alveoli in the upper parts of the lungs because of inadequate pressure head in the pulmonary artery. In an attempt to correlate the presence or absence of the ductus murmur with the presence or absence of shunt flow, it was reasoned that when a large volume of arterial blood was shunted into the pulmonary circulation, there might be more complete perfusion of alveoli in the top of the lung in the upright position. Therefore, the increase in physiologic dead space on assuming this position would not be so marked as might be expected in normal subjects. In this patient, Physiologic dead space

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\text{Tidal volume} \times 100 \text{ increased from 23 per cent in the supine position to 30 per cent in the standing position while the murmur was audible, and from 22 per cent to 49 per cent while the murmur was absent. Although this procedure is probably not sensitive enough to be a practical test for detection of shunts, the results were thought suggestive of an intermittent decrease in pulmonary blood flow associated with the intermittent absence of the ductus murmur.}
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Observations During Surgery

When the chest was open and the mediastinal structures were exposed, a thrill was felt over the ductus arteriosus and the adjacent portion of the left pulmonary artery. Inadvertent rapid infusion of 1 liter of 5 per cent dextrose in water produced no palpable effect on the thrill or on the vital signs. The ductus was about 1 cm. in outside diameter and 1.5 cm. long. It arched superiorly from the aorta and angulated somewhat inferiorly as it entered the pulmonary artery (fig. 3). No unusual nerve to the ductus was found, and mechanical irritation from dissection of adherent structures caused no change in diameter or in the thrill. Slight dorsal displacement of the pulmonary artery by the operator's finger abolished the thrill. This appeared to result from angulation of the ductus so as to close its lumen at the point of curvature several millimeters from its junction with the pulmonary artery.

The patient was not placed in the supine position during surgery. The amount of traction required to abolish the thrill was very slight.
Discussion

The phenomenon of spontaneous intermittent complete disappearance of the typical murmur of patent ductus arteriosus may, in an individual patient, lead to difficulty in arriving at the proper diagnosis. This was apparently true during the earlier years of life in our case. Maneuvers designed to increase cardiac output or elevate systemic pressures might be helpful in bringing out the murmur when it is absent.

In attempting to explain the striking changes in the murmur one is struck by the evidence for intermittency of shunt flow, namely: (1) widened pulse pressure when the murmur was present with distinctly narrower, normal pulse pressures in its absence; (2) physiologic dead space changes suggestive of abnormally increased perfusion of the pulmonary vascular bed when the murmur was audible in contrast to the normal postural changes in its absence; (3) ready abolition of the thrill over the ductus and adjacent left pulmonary artery when gentle dorsal traction was applied to the pulmonary artery. None of the pharmacologic agents made the murmur disappear. Inhalation of a low oxygen mixture resulted in respiratory variations in the murmur rather similar to those occasionally observed during its spontaneous reappearance. This decrease in amplitude with inspiration and increase during expiration of the machinery murmur has been noted in occasional patients with patent ductus arteriosus as well as by Dawes et al. in some of their spontaneously breathing newborn lambs. No hemodynamic data were recorded when these effects were manifest in the present case. It might be suggested, however, that the respiratory variations following the breathing of a low oxygen mixture might have resulted from changes in the aorta to pulmonary artery pressure gradient due to an increase in pulmonary vascular pressure and decreased left ventricular output during inspiration. When these variations occurred spontaneously, inspiratory reductions in left ventricular output alone may have been responsible. The slight reductions in amplitude after amyl nitrite might be explained by mild reductions in flow rate produced by any pressure gradient changes during generalized systemic vasodilatation.

Levine and Harvey stated "in some cases the continuous murmur is intermittent, and its presence and disappearance suggest a fluctuation of the level of pulmonary hypertension." This is considered unlikely in the present instance, for even in the case of primary pulmonary hypertension reported by Gorlin et al., in which acute pulmonary vasoconstriction apparently occurred, the increase in mean pulmonary artery pressure was about 20 mm. Hg. This amount of increase would bring our patient's mean pulmonary artery pressure to less than half her arterial mean pressure, a level probably not high enough to change greatly an established continuous murmur.

Other causes of atypical or absent murmurs such as the presence of associated defects, polycythemia, congestive heart failure appear to have been adequately eliminated.

The cause of the apparent occasional cessation or marked diminution of shunt flow and absence of the murmur in this patient would appear to be mechanical rather than physiologic. In this case acute angulation of the ductus with obliteration of the lumen could have occurred if slight shifts in mediastinal structures can be presumed to have taken place in the intact individual. Quiet rest, particularly in the supine position, might have allowed such shifts to have taken place.

Summary

Transient spontaneous disappearance of the machinery murmur in an otherwise typical case of patent ductus arteriosus unassociated with pulmonary hypertension is described.

Observations of blood pressure changes, physiologic dead space variations in the upright and horizontal postures, and the anatomy of the ductus arteriosus suggested that shunt flow was absent or markedly reduced during the periods in which the murmur was inaudible.
INTERMITTENT MURMUR OF PATENT DUCTUS

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Summario in Interlingua
Es descritbe le transiente o spontanee disparition del murmure de locomotiva in un alteremente typic caso de patente ducto arterioso sin association con hypertension pulmonar.
Observaciones de alterationes in le tension del sanguine, de variationes del spatio morte physiologic in postura erette e horizontal, e le anatonia del ducto arterioso suggereva que shunting de fluxo sanguine esseva absente o marcatemente reducite durante le intervallos quando le murmure esseva inaudibile.

References

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