The Role of Auscultation in the Differentiation of Fallot's Tetralogy from Severe Pulmonary Stenosis with Intact Ventricular Septum and Right-to-Left Interatrial Shunt

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A clinical and phonocardiographic study has been made of the murmurs and heart sounds in the differentiation of Fallot's tetralogy from severe pulmonary stenosis with intact septum and right-to-left interatrial shunt. A striking difference in the behaviour of the systolic murmur was found in the two conditions and this affords a new, simple bedside method of diagnosis. Important differences in the behaviour of the heart sounds were also found, but these are usually more difficult to evaluate clinically and may require special study.

THE value of auscultation in the diagnosis of congenital heart disease has not been sufficiently appreciated. This paper is concerned with the importance of auscultation in the differential diagnosis between Fallot's tetralogy and severe pulmonary stenosis with intact ventricular septum and reversed interatrial shunt. Accurate diagnosis is of great importance. The Blalock-Taussig operation is contraindicated for severe pulmonary stenosis with reversed interatrial shunt, whereas it has proved to be an excellent operation for Fallot's tetralogy.1-6 The diagnosis can generally be made with a reasonable degree of accuracy from consideration of the clinical, radiological and electrocardiographic findings. However, it is often necessary to confirm the diagnosis by means of cardiac catheterisation and angiocardiography. Even with all available methods, certain difficulties arise as discussed elsewhere.6

Any additional, clinical differentiating sign is therefore of value and it is the purpose of this communication to present new physical signs, which will enable accurate diagnosis to be made by careful bedside auscultation. Furthermore, certain clinical and phonocardiographic observations made during the study will be presented.

CLINICAL MATERIAL AND METHODS

All cases accepted for this study presented the common diagnostic problem of pulmonary or infundibular stenosis associated with central cyanosis. This combination usually resolves itself into the diagnosis of Fallot's tetralogy and severe pulmonary stenosis with reversed interatrial shunt. Pulmonary stenosis with rare associations such as transposition of the great vessels, single ventricle and tricuspid atresia were excluded from this study.

Eighteen cases of Fallot's tetralogy and six cases of severe pulmonary stenosis with intact ventricular septum form the basis of this study. All cases had central cyanosis. Cases were accepted only if cardiac catheterisation, selective angiocardiography, intravenous angiocardiography or necropsy proved (a) whether severe pulmonary valvular or infundibular stenosis was present and, (b) whether the ventricular septum was intact or not. If the right-to-left shunt took place in the ventricle through a ventricular septal defect and over-riding aorta, then Fallot's tetralogy was considered proven. If it was established that the ventricular septum was intact Fallot's tetralogy could be excluded. In the latter case a reversed interatrial shunt was assumed to be present though often difficult to prove.8 A complete analysis of the findings in each case is presented elsewhere.4 Necropsy confirmation of the diagnosis was obtained in seven cases.

In addition to full clinical examination, special attention was devoted to auscultation. All cases of pulmonary or infundibular stenosis had a systolic murmur (grade 2 to 5) usually maximal in the second left intercostal space but occasionally in the third and fourth left spaces. Special care was taken
at the site of maximal intensity of the murmur to
determine whether the murmur was loudest in early,
mid or late systole. Even more important, the rela-
tion of the end of the murmur to the second sound
was observed. The presence or absence of splitting
of the second sound was noted. Lastly the first heart
sound and diastole were studied for any abnormality.

The Sanborn Stetho-Cardilette was used to obtain
phonocardiograms in each case. Logarithmic (high
frequency) sound tracings, which give an accurate
graphic representation of human hearing, were
recorded synchronously with a suitable electrocar-
diographic lead, carotid or jugular pulse tracing. The
sound recordings were taken in succession from the
mitral area (XIA), fourth left intercostal space just
to the left of the sternum (4LS), the third left space
(3LS), pulmonary area (PA) and aortic area (AA).
The amplification chosen was purely qualitative,
designed to give the best record for a particular pur-
pose. Thus to record a very soft sound greater
amplification was used. The carotid and jugular
tracings were obtained by placing a crystal micro-
phone directly on the neck, thus avoiding delay from
air conduction.

The components of split first and split second
sounds were identified by determining their respec-
tive relationship to the onset and nadir of the dicrotic
notch of a synchronously recorded right carotid
pulse tracing. Tracings taken at high speed (75 mm.
per second) were essential for accurate study. Since
the dicrotic notch is produced by aortic valve closure,
it can be used to identify the components of a widely
split second sound (figs. 4, 6 to 8). Thus the
onset of the component due to aortic valve closure
precedes the nadir of the notch by a short interval
(average 0.03 second), due to the delay in pulse
wave transmission from the aortic valve to the
carotid artery. It was assumed that the component
occurring after the dicrotic notch could only be due
to pulmonary valve closure. In very wide splitting of
the second heart sound as encountered in severe
pulmonary stenosis, the soft pulmonary component
may be mistaken for a third heart sound. The rela-
tion of this sound to the summit of the "v" wave in
the jugular phlebogram was used to exclude a third
heart sound arising from the right ventricle (figs. 4
and 6). The fact that the soft sound just preceded
the "v" wave indicated that it was produced just
before opening of the tricuspid valve, at the time of
pulmonary valve closure. The occurrence of this
sound only in the pulmonary area and third left
space is in keeping with a soft pulmonary second
sound and unlike a third sound arising in the left
ventricle, which is best heard at the apex or just
internal to it.

Synchronous carotid tracings gave a time relation
between the first sound, added early systolic sounds
and the opening of the aortic valve (figs. 4, 10 and
11).11-13 Delay in pulse wave transmission from aortic
valve to carotid artery was assumed to be given by

the time interval from the onset of aortic component
of the second sound to the nadir of the dicrotic notch.
This interval averaged, as in a previous study,12 0.03
second.

RESULTS

Auscultatory and phonocardiographic find-
ings are shown in tables 1 and 2 and figures
1 and 2.

DISCUSSION

The Second Heart Sound

The second sound is produced by closure of the
aortic and pulmonary valves and asynchronous
closure of these valves results in splitting of the
second sound. Splitting in the pulmonary area is an
almost invariable finding in healthy children and
young adults. The degree of splitting is greatly
influenced by respiration; splitting is widest at the end
of inspiration and narrows or disappears during
expiration. By means of synchronous phonocardiograms
from the mitral, aortic and pulmonary areas,
Leatham and Tower14 showed that the pulmonary
valve normally closes after the aortic valve. This had
also been shown by Wolfik and Margolies in their
studies of bundle branch block, using aortic kymo-
grams and carotid artery tracings.11 Splitting is best
heard in the second and third left intercostal spaces
but not in the mitral and aortic areas because the
normal pulmonary component is not usually con-
ducted to these areas.15-16 Increase in the width of
splitting on inspiration is due to delay of the pul-
monary component. The delayed closure of the
pulmonary valve induced by inspiration has been
attributed not only to the fall in pulmonary artery
diastolic pressure, but also to prolongation of right
ventricular systole from increased filling.16

It has become important during auscultation to
note both the degree of splitting and the intensity of
the two components of the second sound in the
pulmonary area. Splitting may be normal (grade 1 and
2) or abnormally wide (grade 3 and 4).16 A well
known example of abnormal splitting is that often
found in right bundle branch block presumably due
to delayed activation of the right ventricle. Compari-
sion of the intensity of each component also provides
important information. Thus in pulmonary hyper-
tension the pulmonary element is accentuated and is
audible over a wide area even as far as the apex.

We have paid special attention to the pres-
ence or absence of splitting, and to the degree
of reduction of intensity of the pulmonary
component in cases of pure pulmonary stenosis
of differing severity and in cases of Fallot’s
tetralogy. Pulmonary valvular stenosis and
infundibular stenosis, unlike aortic stenosis,
Table 1.—Auscultatory Findings in Six Patients with Severe Pulmonary Stenosis with Intact Ventricular Septum and Right-to-Left Interventricular Shunt

<table>
<thead>
<tr>
<th>No.</th>
<th>Case, age, sex</th>
<th>Central cyanosis (1–4)</th>
<th>Disability (1–4)</th>
<th>Site of max. intensity</th>
<th>Grade (1–6)</th>
<th>Relation to 2nd sound</th>
<th>Systolic murmur (at site of max. intensity)</th>
<th>2nd sound (at site of max. intensity of SM.)</th>
<th>1st sound</th>
<th>Atrial sound</th>
<th>Diast. murmur</th>
<th>Phonocardiogram</th>
<th>Site of stenosis</th>
<th>Proof</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>E. B., 49, F</td>
<td>2</td>
<td>3</td>
<td>PA</td>
<td>4</td>
<td>Single</td>
<td>Long SM. Stops before soft P2. As not heard.</td>
<td>Single Very soft on exp. only. Max. in PA.</td>
<td>Loud on inap.</td>
<td>0</td>
<td>As buried in prolonged SM which stops before soft P2. Width of split about 0.13 sec. Loud first sound on expir.; loud atric. sound on inap.</td>
<td>Valvular</td>
<td>Neopaty</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>S. J., 9, F</td>
<td>1</td>
<td>1</td>
<td>4LS</td>
<td>5</td>
<td>0</td>
<td>Long SM. A1 &amp; P2 inaudible.</td>
<td>0</td>
<td>N</td>
<td>0</td>
<td>As buried in prolonged SM. No P1 recorded. Normal A1 at MA &amp; AA.</td>
<td>Valvular</td>
<td>Cath. Angio. Valvotomy</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>P. P., 42, M</td>
<td>1</td>
<td>2</td>
<td>PA</td>
<td>4</td>
<td>Split (Very wide.)</td>
<td>Long SM. Stops before soft P2. As just audible.</td>
<td>As N P2 very soft on exp. only. Max. 3LS.</td>
<td>Loud on inap.</td>
<td>0</td>
<td>As buried by prolonged SM, which stops before soft P2. Width of split ca. 0.11 sec. Loud first sound on exp., loud atric. sound on inap.</td>
<td>Valvular</td>
<td>Cath. Angio. Valvotomy</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>S. D., 10, M</td>
<td>1</td>
<td>1</td>
<td>PA</td>
<td>5</td>
<td>0</td>
<td>Long SM. A1 &amp; P2 inaudible.</td>
<td>0</td>
<td>Loud and sharp.</td>
<td>0</td>
<td>As buried by prolonged SM which stops before soft P2. Width of split ca. 0.11 sec.</td>
<td>Valvular</td>
<td>Cath. Sel. angio (RV.)</td>
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</tbody>
</table>

A1 = Aortic component of second sound. P2 = pulmonary component of second sound. Sel. angio (RV.) = selective angiocardiogram from right ventricle.
result in a fall in pressure distal to the obstruction and in consequence the second or pulmonary component of the second sound becomes soft or inaudible depending upon the degree of stenosis. Furthermore, the low pulmonary diastolic pressure and probable prolongation of the right ventricular systole results in delayed closure of the pulmonary valve so that wide splitting of the second sound is heard or recorded provided the pulmonary component is loud enough.

The Second Sound in Severe Pulmonary Stenosis with Closed Ventricular Septum. It is easier to understand the findings in severe pulmonary stenosis if one first considers the changes that occur in mild pulmonary stenosis. In most cases of mild pulmonary stenosis, as defined by Abrahams and Wood, we heard and recorded a normal to widely split second sound with a normal to soft pulmonary component (fig. 3). The pulmonary systolic murmur in all cases filled systole extending up to the first or aortic component of the split second sound. Even in cases of moderate pulmonary stenosis it was often possible to hear wide splitting of the second sound but the pulmonary component was always abnormally soft, thus confirming the previous observations of Leatham. An exaggeration of these findings might be anticipated in severe pulmonary valvular stenosis.

In severe pulmonary stenosis clinical analysis of the altered second sound is difficult. In three of the 6 cases the harsh prolonged pulmonary systolic murmur stopped before an extremely soft sound, but in the remaining three no pulmonary component could be heard. In one case only (case 5, P. P.) was very wide splitting (grade 4) appreciated by the ear. In this case the aortic component was not completely obscured by the murmur. The phonocardiogram with synchronous carotid and jugular tracings were essential in analysing these changes. In each case the systolic murmur in the pulmonary area was so prolonged that it had extended beyond the first or aortic component of a widely split second sound (figs. 4 and 8). In fact the aortic sound was completely buried in the murmur in all except one case. The buried sound could be identified by the dicrotic notch of the carotid tracing and also by sound tracings taken at the apex or fourth left space where a normal aortic sound was less obscured by the systolic murmur (fig. 4). The systolic murmur ended before a very soft pulmonary second sound which was recorded in four out of six cases, thus proving that very wide splitting was in fact present in severe pulmonary stenosis. In the remaining two cases the second sound was presumably so soft that even considerable amplification failed to reveal its presence.

Thus in severe pulmonary stenosis with intact septum, although the phonocardiogram demonstrates very wide splitting of the second sound and may therefore be useful as a special test, clinical auscultation at the site of maximal intensity merely reveals a harsh prolonged systolic murmur which may be followed by a very soft pulmonary second sound. However, auscultation in the mitral area and fourth left space where the murmur is less intense reveals an aortic second sound of normal intensity.

The Second Sound in Fallot's Tetralogy. In Fallot's tetralogy, where the severity of the stenosis is of an order comparable to severe pulmonary stenosis with intact septum, a similar alteration of the second sound might be anticipated. This is, in fact, not the case. In Fallot's tetralogy the second sound is often abnormally loud and thus usually palpable (table 2). This may be due to the fact that the aortic valve is brought closer to the anterior chest wall by the dextroposition of the aortic root. Furthermore, the systolic murmur finishes or diminishes markedly before the aortic second sound (vide infra) thereby contributing to the striking clarity of the second sound in Fallot's tetralogy. Phonocardiograms confirmed these clinical findings in all cases (figs. 2 and 5). Comparison of figure 4 with figure 5 shows how the aortic second sound is obscured by the prolonged systolic murmur in severe pulmonary stenosis with intact septum whereas in Fallot's tetralogy the loud aortic second sound stands well to the right of a shorter pulmonary systolic murmur.

The problem of the pulmonary second sound in Fallot's tetralogy is of considerable academic
### Table 2.—Auscultatory Findings in 18 Patients with Fallot's Tetralogy

<table>
<thead>
<tr>
<th>No.</th>
<th>Case, age, sex</th>
<th>Central cyanosis (1–4)</th>
<th>Disability (1–4)</th>
<th>Site of max. intensity</th>
<th>Grade</th>
<th>Relation to 2nd sound</th>
<th>Systolic murmur (at site of max. intensity)</th>
<th>2nd sound (at site of max. intensity of SM.)</th>
<th>Intensity</th>
<th>1st sound</th>
<th>Atrial sound</th>
<th>Diast. murmur</th>
<th>Phonocardiogram</th>
<th>Site of stenosis</th>
<th>Proof</th>
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<tr>
<td>9</td>
<td>E. W., 19, M</td>
<td>3</td>
<td>1</td>
<td>PA</td>
<td>3</td>
<td>Single</td>
<td>SM stops before A2.</td>
<td>Single</td>
<td>Loud (pal-pable)</td>
<td>N</td>
<td>0</td>
<td>0</td>
<td>SM stops just before loud A2. No P.</td>
<td>Infundib.</td>
<td>Neeropay</td>
</tr>
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<td>12</td>
<td>E. K., 10, M</td>
<td>2 (Pre-op 3)</td>
<td>3 (Pre-op 4)</td>
<td>3LS</td>
<td>2</td>
<td>Single</td>
<td>Short SM. Stops well before A2.</td>
<td>Single</td>
<td>Loud (pal-pable)</td>
<td>Aortic early systolic sound at all areas.</td>
<td>0</td>
<td>0</td>
<td>Short SM. Stops long before loud A2. No P. Split 1st sound plus aortic early systolic sound.</td>
<td>Infundib.</td>
<td>Cath. Blalock-Taussig op.</td>
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<tr>
<td>No</td>
<td>Name</td>
<td>Age</td>
<td>Sex</td>
<td>Status</td>
<td>Findings</td>
<td>Condition</td>
<td>Follow-Up</td>
<td></td>
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Auscultation in Diagnosis of Congenital Heart Disease

Fig. 1. Six cases of Severe Pulmonary Stenosis with Intact Ventricular Septum and Right-to-Left Interventricular Shunt. In all the phonocardiograms were taken at the site of maximal intensity of the murmurs. In each case the systolic murmur (SM) is intense and so prolonged that it extends beyond and drowns the normal aortic component (A) of the very widely split second sound (2, 2'). It stops before the delayed diminutive pulmonary component (P) when recorded. With the exception of case 4, which was recorded after a partially successful pulmonary valvotomy, the approximate position of the buried aortic component is indicated by the dicrotic notch of the synchronously recorded carotid pulse. Black dots have been used to indicate the limits of the sound vibrations in this and subsequent figures.

interest. It is generally stated that a pure single second sound occurs in Fallot’s tetralogy. While this is usually true our observations prove that in some cases the pulmonary second sound follows the aortic second sound and we believe that splitting of the second sound in Fallot’s tetralogy would be elicited more often were it not for the fact that the pulmonary valve closure is so quiet. In one remarkable case (case 17, S. P.) of proven Fallot’s tetralogy, wide splitting of the second heart sound was easily detected clinically (fig. 6) and gave rise to considerable diagnostic difficulties. The unusually high pulmonary artery pressure found in this case seemed the best explanation of the audible pulmonary second sound. In case 10 (C. G.) careful auscultation two months after a successful Blalock-Taussig operation revealed definite splitting of the second heart sound whereas prior to operation only a single second sound was heard and recorded (fig. 7).

The appearance of the soft pulmonary second sound after the operation suggested that the pulmonary artery pressure had risen above a critical level necessary for production of audible valve closure. In two further cases (case 8, W. K., and case 21, L. K.) a definite diminutive pulmonary second sound was recorded but not heard. This evidence suggests that the usual single second sound in Fallot’s tetralogy is not due to synchronous closure of the aortic and pulmonary valves. It is, in fact, due to aortic valve closure alone, which is followed by pulmonary valve closure, presumably inaudible because of the low pulmonary diastolic pressure.

Although the series is small, the width of splitting in Fallot’s tetralogy appears to be considerably less than that in severe pulmonary stenosis with intact septum (figs. 4, 6, 7 and 8). In both conditions there is a low pulmonary diastolic pressure which is an
FIG. 2. *Eighteen Cases of Fallot's Tetralogy.* In all cases the phonocardiograms were taken at the site of maximal intensity of the murmur. In each case the murmur commences soon after the first sound, reaches maximal intensity by mid-systole, and then diminishes markedly, usually ending before the single, loud aortic component of the second sound which is not in any way obscured by the murmur. In the four most severe cases (cases 12, 13, 15 and 22) the systolic murmur was soft, early and short.
Fig. 3. Synchronous phonocardiogram (PCG) at the pulmonary area (PA) and lead II (75 mm./sec.), from a case of mild pulmonary valve stenosis with intact septum (RVP 30/0; PAP 10/4 mm. Hg). The loud systolic murmur (SM) fills systole, extending up to, but not obscuring, the first or aortic component (A) of the well split second sound. The second or pulmonary component (P) is moderately reduced in intensity.

important common factor in the production of the split sound. It is suggested that the greater delay of the pulmonary component in severe pulmonary stenosis with intact septum is due to the additional factor of prolongation of right ventricular systole (vide infra).

The Systolic Murmur

In all cases studied a systolic murmur was present. The murmur was usually harsh and loud (grade 3 to 5) in which case its intensity was of no help in differentiating the two conditions. However, in four cases of Fallot's tetralogy (cases 12, 13, 15 and 22), all severe, the systolic murmur was soft (grade 1 or 2). On this basis alone Fallot's tetralogy was favored as in all cases of severe pulmonary stenosis with intact ventricular septum the murmur was loud.* The site of maximal intensity, however, was of no real help in the differential diagnosis. Where the stenosis was valvular in both conditions, the murmur was maximal at the second and third left intercostal spaces (with the exception of case 2, S. J.). In infundibular stenosis the murmur was as often maximal in the second as in third and fourth intercostal spaces. A murmur maximal in the fourth space favored infundibular stenosis and hence Fallot's tetralogy because infundibular stenosis occurs so much more commonly in this condition. However, in one case of valvular stenosis with intact ventricular septum (S. J.), the murmur was maximal in the fourth intercostal space.

Previous studies of the systolic murmur have failed to reveal any characteristic distinguishing feature.10, 16-20 We agree that the loudness and the site of maximal intensity of the murmur is usually of no real help in the differential diagnosis of Fallot's tetralogy from severe pulmonary stenosis with intact septum. However, we have found that there is a striking difference in the two murmurs, if attention is directed to the duration of the systolic murmur in relation to the second sound and to the position in systole of maximal intensity. Our findings have led us to believe that an accurate diagnosis can be made by careful bedside auscultation.

The Systolic Murmur in Severe Pulmonary Stenosis with Intact Ventricular Septum. Auscultation at the site of maximal intensity of the murmur reveals a prolonged systolic murmur. The aortic component of the second sound is rarely heard, being drowned by the crescendo of the murmur (figs. 1, 4, 8). The murmur stops before the delayed diminutive pulmonary component which may occasionally be heard by very careful auscultation in the pulmonary area. In the mitral area where the systolic murmur is much less intense, the aortic second sound, of normal intensity, is not drowned and is therefore usually clearly audible (fig. 4).

The Systolic Murmur in Fallot's Tetralogy. In Fallot's tetralogy the behaviour of the pulmonary systolic murmur is quite different. However loud or soft, the systolic murmur at the site of maximal intensity, is much less prolonged than in severe pulmonary stenosis with intact ventricular septum. It commences soon after the first sound, reaches maximal intensity by mid-systole and then diminishes markedly, usually ending before the single loud aortic component of the second sound (figs. 2, 5, 6, 7). The phonocardiogram may record small rapidly diminishing vibrations right up to the second sound, but these are

*We have since encountered one case of unusually severe pulmonary stenosis in heart failure in whom the systolic murmur was grade 2 only.
Fig. 4. Case 5 (P. P.) Severe Pulmonary Stenosis with Intact Septum. The systolic murmur at the site of maximal intensity (PA) is so prolonged that it extends beyond the first or aortic component (A) of the very widely split second sound (2, 2'). The aortic sound is, in fact, completely buried in the murmur. The buried sound can be identified by the dicrotic notch of the carotid tracing (CAR) and by the PICG taken at the mitral area (MA) where the normal aortic sound (A) is not obscured by the systolic murmur. The systolic murmur ends before the soft pulmonary component (P). The latter is identified by the fact that it just precedes the "V" wave of the jugular venous tracing (JUG) indicating that it is produced just before opening of the tricuspid valve, at the time of pulmonary valve closure. The tracing also records the extremely intense "second component" (1') of the first sound maximal in the PA commencing 0.01 second before the onset of ejection into the aorta (a delay of 0.03 second in pulse wave transmission from the aortic valve to the carotid artery has been assumed). As the heart rate was ident-

not detected by the ear, the murmur appearing to end well before the loud second sound.

As far as we are aware this difference in the behavior of the pulmonary systolic murmur in the two conditions has not been hitherto described.

**Comment**

The striking difference in the duration of the systolic murmur can probably be related

ideal, the tracings as mounted, here and elsewhere, are virtually synchronous.
Figure 6. Case 17 (S. P.) Fallot's Tetralogy. The systolic murmur commences soon after the first sound and ends before the loud aortic component (A) of the second sound which is identified by the dicrotic notch of the carotid tracing. The unusually loud pulmonary component (P) is identified by the fact that it just precedes the "V" wave of the jugular venous tracing. The width of splitting measures 0.08 second as compared with 0.11 second in figure 4 and ca. 0.14 second in figure 8.

Figure 7. Case 10 (C. G.) Fallot's Tetralogy. (a) The systolic murmur reaches maximal intensity by mid-systole and diminishes markedly before the loud aortic component of the single second sound. (b) Two months after a Blalock-Taussig operation the pulmonary component (P) of the second sound became just audible giving rise to splitting of the second sound (2, 2') measuring 0.08 second and assists it in generating such high pressures.30-40 A considerable difference in the amplitude and duration of the pressure curves in the right and left ventricles must occur. We have shown that the second sound is widely split due to marked delay of the very soft pulmonary component. The delayed closure of the pulmonary valve is due not only to the low pulmonary diastolic pressure, but also to prolongation of the ejection phase of right ventricular systole as compared with that of the left ventricle. Flow will continue across the stenotic pulmonary valve as long as a pressure gradient exists between the right
ventricle and the pulmonary artery. Thus when the aortic valve closes at the end of the normal or reduced left ventricular systolic active flow still continues across the pulmonary valve. Since the intensity of the systolic murmur is probably proportional to the speed and volume of flow, other factors such as degree of stenosis and viscosity being equal, it is not surprising to find the pulmonary systolic murmur continuing beyond the aortic second sound (diagram 1).

In Fallot’s tetralogy, unlike severe pulmonary stenosis with intact ventricular septum, the ventricular septal defect and the over-riding aorta both act as a safety valve to the right ventricle; no matter how severe the pulmonary or infundibular stenosis, the right ventricular systolic pressure can never significantly exceed that in the aorta and left ventricle. Thus clinical evidence of right ventricular and right atrial hypertrophy is much less. This is shown by the absence of a giant “a” wave in the venous pulse, the slight lift over the right ventricle and the absence of marked right ventricular enlargement radiologically. In Fallot’s tetralogy the right and left ventricles virtually function as a single ventricle from which blood is discharged into two competing orifices, namely the aortic with normal systemic peripheral resistance* and a stenotic pulmonary valve or infundibulum with a much higher resistance. The ventricular pressure curve in both ventricles is conditioned by the lesser resistance offered by the aorta and hence adaptation to the severe pulmonary stenosis is much less effective than in pulmonary stenosis with an intact septum. Thus effective systole cannot be more prolonged in the right ventricle than the left because by the time the aortic valves close the ventricles have virtually discharged their contents. This is well illustrated by angiocardiography which reveals rapid discharge of diodone from the right ventricle into the aorta in Fallot’s tetralogy contrasting sharply with the delayed emptying of the right ventricle seen in severe pulmonary stenosis with intact septum. Maximal flow across the stenotic valve or chamber occurs early in systole at the time of

* This has not been measured by us but the absence of hypertension in our cases of Fallot’s tetralogy at least indicates a normal systemic peripheral resistance.
maximal flow into the aorta; during the phase of reduced ejection from the ventricles into the aorta, flow into the pulmonary artery is likewise reduced. By the time the aortic valve closes flow into the pulmonary artery has virtually ceased being entirely dependent on the rapidly diminishing pressure gradient between the right ventricle and pulmonary artery and the little available residual blood.

It is therefore not surprising that the pulmonary systolic murmur* in Fallot's tetralogy attains maximal loudness by mid-systole and then diminishes rapidly, usually ending before the aortic second sound. Variation in the intensity and duration of the murmur does, however, occur. In our four most disabled patients (cases 12, 13, 15 and 22, in three of whom extreme stenosis was proven at necropsy) the murmur was soft and short reaching a crescendo well before mid-systole and ending long before the aortic second sound (figs. 2, 12 and 10). A short murmur thus appears to indicate severe stenosis. This inference is not unreasonable for the severer the stenosis the less the flow across the pulmonary valve. Unlike pulmonary stenosis with intact septum, the systemic peripheral resistance and not the pulmonary stenosis determines the level of the right ventricular systolic pressure. This is grossly inadequate in extreme pulmonary stenosis and thus flow sufficient to produce a murmur is limited to the early ejection phase of the right ventricle. In milder cases of Fallot's tetralogy the murmur appears to be longer (fig. 12) but we have not yet en-

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countered any case in which the murmur obscured the second sound.†

Auscultatory Observations of Lesser Diagnostic Value

Diastolic Murmurs. No case had pulmonary or aortic incompetence. In one case of severe pulmonary stenosis with intact ventricular septum (case 3, J. F.), a presystolic murmur was well heard in the second, third and fourth intercostal spaces, being loudest in the pulmonary area. It was not heard at the apex. The murmur was preceded by a loud atrial sound also best heard at the pulmonary area (fig. 8). The murmur occurred after the onset of right atrial contraction which was unusually powerful as revealed by the giant "a" wave (fig. 8). Cardiac catheterisation showed a right atrial systolic pressure of considerable magnitude transmitted into the left atrial and pulmonary veins as reported elsewhere.6 Abrahams and Wood8 have encountered in severe pulmonary stenosis a similar presystolic murmur which they attributed to flow across a patent foramen ovale shown at necropsy. In our case flow across the pulmonary valve produced by the powerful right atrial contraction may be an alternative explanation.

The Atrial Sound. In three cases of severe pulmonary stenosis with intact ventricular septum (cases 1, 3 and 5), a singularly loud

† Unusual prolongation of the systolic murmur in Fallot's tetralogy may occasionally occur. The second sound, however, is still not obscured by the murmur. In our only case of proven Fallot's tetralogy where this occurred, coarctation of the aorta was also present, causing a systemic blood pressure of 170/80 mm. Hg. It is probable that the unusually high right ventricular systolic pressure conditioned by the high aortic resistance permitted vigorous flow to continue through the stenosed pulmonary valve for a longer duration than the usual case of Fallot's tetralogy. This is supported by the fact that the patient's cyanosis had disappeared clinically during observation over several years, and his effort tolerance had increased. Furthermore, the pulmonary artery pressure was unusually high and the second sound was widely split with a very soft but audible pulmonary component. Being complicated by coarctation the case has not been included in this series but is discussed elsewhere.4
An extremely loud first sound appears suddenly during expiration only to disappear completely during inspiration (INSP.). It occurs just after the first sound and before the onset of the carotid pulse. The marked effect of respiration on the atrial (A) is recorded, being loud during inspiration and absent during expiration. The characteristic systolic murmur of pulmonary stenosis with intact septum is also shown.

The First Heart Sound. Several points of interest emerged from the study of the first sound which require further investigation and may prove to be of additional diagnostic value.

In Fallot’s tetralogy the first sound was usually normal, being either single or closely split. In four cases it was apparently “widely split”. The second component, however, was not attributed to the second component of a physiologically split first sound but to an aortic early systolic sound because of the wide transmission to all areas including the aortic area and the marked degree of splitting. Furthermore, the phonocardiogram proved the separate identity of this added sound by showing normal splitting of the first sound preceding the aortic sound (fig. 10). Moreover, this sound occurred about 0.04 second after ejection into the aorta, a delay of 0.03 second in pulse wave transmission from aortic valve to carotid artery being assumed. The aorta is dextroposed and nearer the chest wall than normal. It is dilated and receives a greater

auricular sound was heard and recorded in the second, third and fourth intercostal spaces but not at the apex (figs. 8 and 9). The intensity was markedly influenced by the phase of respiration, being loud during inspiration and soft during expiration (fig. 9). This sound was attributed to vibrations occurring in the right ventricle or pulmonary valve produced by the powerful atrial systolic filling wave.

In no case of Fallot’s tetralogy was an atrial sound heard. This is in keeping with the concept already discussed that the right ventricle and right auricle in Fallot’s tetralogy are under far less stress than in severe pulmonary stenosis with intact ventricular septum. It is concluded that an audible atrial sound favours the diagnosis of severe pulmonary stenosis with intact septum.

Fig. 9. Case 1 (E. B.). Severe Pulmonary Stenosis with Intact Septum. (a) Synchronous phonocardiogram at 3 LS and carotid tracing at 75 mm./sec. (b) synchronous phonocardiogram at 3 LS and lead III at 25 mm./sec.
volume of blood than normal, being the main exit of blood from both ventricles. These factors may be responsible for this audible ejection sound. The presence of this sound appears to be an indication of the severity of the lesion. The more severe the pulmonary stenosis the greater the volume of blood ejected into the aorta, and hence the larger the aorta. This combination favours the production of an audible ejection sound. This sound was only present in our four most disabled cases (cases 12, 13, 15 and 22). Furthermore, in two cases of pulmonary atresia, where the output of both ventricles was discharged into a single dilated “pseudotruncus arteriosus”, a striking aortic systolic sound was heard (fig. 11). Although the aortic early systolic sound has been described in a variety of conditions affecting the aorta and aortic valves, its occurrence in Fallot’s tetralogy is much less well known and in pulmonary atresia it has not been described before.

In severe pulmonary stenosis with intact septum the first sound was usually normal. However, in two cases of this series a remarkable abnormality with striking respiratory variation was found (figs. 4 and 9). An extremely loud, palpable, click-like first sound would suddenly appear during expiration only to disappear completely during inspiration. It occurred just after the first sound, giving the impression of close splitting. Phonocardiograms showed that this sound was present only in expiration and was synchronous with the second component of a closely split first sound occurring before the onset of the carotid pulse (figs. 4 and 9). Clinically, it was quite unlike the second component of a normally split first sound both in intensity and in that it was maximal at the pulmonary area and not the mitral area or fourth intercostal space.

This was confirmed in one of the two cases (case 1, E. B.) when the heart was palpated at operation and the sound appeared to originate in region of the pulmonary valve. It thus appears to be related to the phase of ejection from the right ventricle and not to auriculo-ventricular valve closure. Because of the great shortening of the isometric contraction phase
Fig. 12. The Duration of the Systolic Murmur in Relation to the Severity of Fallot's Tetralogy. In pulmonary atresia there is usually no systolic murmur. In Fallot's tetralogy with extreme stenosis the murmur is short, soft and confined to early systole. In both these conditions an aortic early systolic sound (X) is usually heard. In the majority of cases of the tetralogy the systolic murmur is loud, reaches its crescendo by midsystole and diminishes rapidly before the loud aortic second sound. In mild cases of the tetralogy the murmur appears to be of longer duration, as shown in the tracing taken six months after a successful valvotomy. The murmur extends up to the aortic second sound (A) but does not obscure it. The pulmonary second sound (P) has become audible, producing wide splitting (0.09 second), indicating Fallot's tetralogy with mild stenosis. (The pulmonary valvotomy has presumably produced a rise in the pulmonary diastolic pressure.)

of the right ventricle, due to the very low pulmonary diastolic pressure, this ejection sound may be almost superimposed on the tricuspid component of the first heart sound. Like the pulmonary early systolic sound in dilatation of the pulmonary artery, this sound is also related to the phase of early ejection from the right ventricle into the pulmonary artery. The mode of production, however, is possibly different. Audible vibrations may be set up by the sudden tautening of the stenotic pulmonary valve bulging into the pulmonary artery at the moment of ejection into the pulmonary artery. This sign is not confined to severe pulmonary stenosis but has also been encountered in moderate and mild stenosis. It was first described by Petit in 1902 and by several workers since. We have not encountered this sign in Fallot's tetralogy.

Summary

1. The differentiation of Fallot's tetralogy from severe pulmonary stenosis with intact ventricular septum and reversed interatrial shunt is important because the surgical treatment is different. While the diagnosis can generally be made on clinical grounds alone, it is usually necessary to confirm the diagnosis by special investigations.

2. A clinical and phonocardiographic study of the heart sounds and murmurs has been made in six cases of severe pulmonary stenosis with intact septum and in 18 cases of Fallot's tetralogy.

3. A striking difference in the behaviour of the systolic murmur at the site of maximal intensity was found in the two conditions, if attention was directed to the duration of the systolic murmur in relation to the second heart sound and to the position in systole of its maximal intensity. In Fallot's tetralogy the systolic murmur, however soft or loud, starts soon after the first sound, reaches maximal intensity by mid-systole and then diminishes markedly, usually ending before the single loud, often palpable, aortic component of the second sound. By contrast, in severe pulmonary stenosis with intact ventricular septum, the systolic murmur is so prolonged, that it extends beyond and drowns the normal aortic component of the very widely split second sound. It stops before the delayed, diminutive pulmonary component which may or may not be audible.

4. This significant difference in the systolic murmur so clearly shown by phonocardiography is readily appreciated by the ear and affords a new simple bedside method of diagnosis.

5. Phonocardiographic studies revealed
further points of interest. In severe pulmonary stenosis with intact septum the second heart sound is very widely split due to a diminutive pulmonary second sound being widely separated from the normal aortic component. In Fallot's tetralogy the second sound is usually single because the loud aortic sound is followed by an inaudible pulmonary sound. However, if the pulmonary artery pressure is sufficiently high, this delayed pulmonary second sound becomes recordable and even audible. The mechanism underlying these findings is fully discussed.

6. A presystolic murmur loudest in the left parasternal region was heard in a case of severe pulmonary stenosis with intact ventricular septum and reversed interauricular shunt. It was attributed to flow through the atrial septum or the stenosed pulmonary valve during atrial systole.

7. A loud atrial sound was heard in three cases of severe pulmonary stenosis with intact septum but not in Fallot's tetralogy.

8. In four severe cases of Fallot's tetralogy in whom the murmur was not intense, an early systolic sound was heard which caused "wide splitting" of the first sound. In two cases of severe pulmonary stenosis with intact ventricular septum close splitting of the first sound was heard with an intense clicking second component. The mechanism underlying these findings is discussed.

**SUMMARIO IN INTERLINGUA**

1. Le differentiation de tetralogia de Fallot ab sever stenosis pulmonar con intacte septo ventricular e revertite derivation interatrial es importante proque le tractamento chirurgic differe in le duo casos. Ben que le diagnoste es generalmente facibile super le base de datos clinic, il es usualmente necessari verificare le diagnoste per investigationes special.

2. Esseva executate un studio clinic e phonocardiographic del sonos e murmures cardiac in sex casos de sever stenosis pulmonar con intacte septo e in dece-octo casos de tetralogia de Fallot.

3. Esseva trovate un frappante differencia inter le duo conditiones in le murmure systolic al sito de intensitate maximal. Iste differencia esseva manifeste in le relation inter le duration del murmure systolic e le secunde sono cardiac e in le puncto del systole ubi le murmure attingeva su intensitate maximal. In casos de tetralogia de Fallot le murmure systolic—sin reguardo a si illo es forte o basse—comencia tosto post le prime sono; illo attinge su intensitate maximal in mediysystole; e tunc illo diminue marcatamente e se termina usualmente ante le unie e forte e frequentemente palpabile component aortic del secunde sono. Per contrasto con isto, in casos de sever stenosis pulmonar con intacte septo ventricular, le murmure systolic es si prolongate che illo ultrapassa e inunda le normal component aortic del secunde sono che alora es nettemente dividite. Illo se termina ante le retardate e minime component pulmonar que a vices es audibile e a vices non.

4. Iste significative differencia in le murmure systolic, que se demonstra si clarnemente per medios phonocardiographic, es facilmente perceptibile per le aure e representa un nove e simple metodo de diagnose clinic.

5. Studios phonocardiographic revelava altere punctos de interesse. In casos de sever stenosis pulmonar con intacte septo, le secunde sono es dividite a longe intervallo in consequentia del facto que un minime secunde sono pulmonar es nettemente separate ab le normal componente aortic. In tetralogia de Fallot le secunde sono es generalmente non-dividite proque le forte sono aortic es sequite per un inaudibile sono pulmonar. Nonobstante, si le pression pulmonar arterial es sufficientemente alte, le retardate secunde sono pulmonar deveni registrabile e mesmo audibile. Le mechanismo al base de iste constatationes es discutite.

6. Un murmure presystolic que habeva su intensitate maximal in le region sinistro-para-sternal esseva audite in un caso de sever stenosis pulmonar con intacte septo ventricular e revertite derivation interauricular. Illo esseva attribuite a un fluxo a transverso le septo auricular o a un stenosis del valvula pulmonar durante le systole auricular.

7. Un forte sono auricular esseva audite in tres casos de sever stenosis pulmonar con intacte septo sed non in tetralogia de Fallot.
8. In quatro sever casos de tetralogia de Fallot le murmure non esseva intenso, sed il se audiva un prompte sono systolic que causava un division a grande intervallo del prime sono. In duo casos de seve stenosis pulmonar con intachte septo ventricular, un division a breve intervallo esseva audite in le prime sono. Le secunde componente representava un intenso clie. Le mechanismo al base de iste constatazioni es discutite.

Addendum

Since this paper was submitted we have encountered three further cases of severe pulmonary stenosis with intact ventricular septum and right-to-left interatrial shunt and eight cases of Fallot’s tetralogy. In each case a correct diagnosis was made from auscultation alone. Furthermore, in Fallot’s tetralogy it was usually possible to assess whether the stenosis was extreme, average or mild by paying careful attention to the auscultatory features discussed above (fig. 12). These cases are discussed elsewhere.

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The Role of Auscultation in the Differentiation of Fallot's Tetralogy from Severe Pulmonary Stenosis with Intact Ventricular Septum and Right-to-Left Interatrial Shunt

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