Internal Mammary Artery-to-Pulmonary Artery Fistulas

Case Report and Review of the Literature

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SUMMARY A case of internal mammary artery-to-pulmonary artery fistula is presented, and all previously reported cases are reviewed. This represents a rare cause of a continuous precordial murmur that may be confused clinically with a patent ductus arteriosus. Patients are usually asymptomatic, and routine diagnostic tests may be of little help in establishing a diagnosis. Selective internal mammary angiography is the procedure of choice for confirmation and localization. The question of advisability of surgery for these patients remains unanswered.

ARTERIOVENOUS FISTULAS of the internal mammary artery are rare.\textsuperscript{1-3} In a review of the 10-year experience with arteriovenous fistulas at the Mayo Clinic between 1955 and 1965, only two of 181 cases involved chest-wall vessels.\textsuperscript{4} Rarely, a connection may exist between an internal mammary artery and the pulmonary arteries, clinically mimicking a patent ductus arteriosus because of the resultant continuous murmur in the parasternal region.\textsuperscript{5}

We have been able to locate 10 well-described cases of communication between an internal mammary artery and a pulmonary artery in the world literature, and we describe here the eleventh such case. This is only the second case in which the communication was to the main pulmonary artery rather than to one of its branches.

Case Report

An 18-year-old Caucasian female was referred to the Mayo Clinic in March 1979 because of a heart murmur. The murmur was noted in 1977 by her family physician during a prenatal checkup. The murmur was again noted in early 1979, and she was referred to this institution for cardiology consultation.

On evaluation here, she denied having symptoms of heart disease. The systems review revealed only morning headaches during pregnancy. The patient had no history of surgery or trauma, and there was no family history of heart murmurs or congenital anomalies.

On physical examination, her pulse was 84 and regular, and her blood pressure was 108/80 mm Hg in the right arm and 120/90 mm Hg in the left arm. There was no cyanosis or clubbing. No skeletal or other anomalies were noted. The heart was not palpably enlarged, and there was no palpable thrill. The heart sounds were normal, without gallop rhythm.

There was a grade 2/6 murmur, heard best at the left sternal border at the second interspace; this was present throughout systole and continued through the second heart sound into diastole.

A posteroanterior chest film (fig. 1) and a resting ECG were normal. The laboratory data, including a complete blood count and blood chemistries, were also normal. An M-mode echocardiogram revealed normal valvular motion and chamber sizes. Exercise electrocardiography was not done.

Right- and left-heart catheterization was carried out on March 21, 1979 (table 1). Hemodynamic and oximetric data revealed no abnormalities. Double-sampling indocyanine green dye-dilution curves were done with injection of the aortic root and sampling of the right-sided chambers. A small (less than 5.0%) left-to-right shunt at the pulmonary artery level was found. An aortic root angiogram and a selective left internal mammary artery angiogram showed an anomalous connection between the left internal mammary artery and the main pulmonary artery (fig. 2).

No surgery was advised. The patient was instructed in prophylactic precautions for subacute bacterial endocarditis, but no restrictions were placed on her activities.

Discussion

The first report of a connection between the internal mammary vessels and a pulmonary artery was made by Burchell and Clagett\textsuperscript{4} from this institution in 1947. In that patient, a pulmonary arteriovenous fistula was considered to be the initial anomaly; the connection from the internal mammary artery was believed to be a secondary, acquired condition.

Arteriovenous fistulas of the internal mammary artery may be divided into three groups on the basis of their anatomic connections.\textsuperscript{1} Fistulas derived from the proximal portion of the internal mammary artery connect with the internal mammary vein, vena cava or innominate vein, those from the middle third of the internal mammary artery connect with the pulmonary circulation, and those from the distal third connect to the vitelline venous system.\textsuperscript{7} Other shunts from systemic arteries to the pulmonary artery may originate from intercostal, bronchial, epicardial, pericardio-
TABLE 1. Catheterization Data

<table>
<thead>
<tr>
<th>Position</th>
<th>Pressure (mm Hg)</th>
<th>Saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td>—</td>
<td>77</td>
</tr>
<tr>
<td>IVC</td>
<td>—</td>
<td>76</td>
</tr>
<tr>
<td>RA</td>
<td>12/6</td>
<td>74</td>
</tr>
<tr>
<td>RV</td>
<td>22/0-8</td>
<td>73</td>
</tr>
<tr>
<td>PA</td>
<td>21/10</td>
<td>—</td>
</tr>
<tr>
<td>LPA</td>
<td>22/9</td>
<td>74</td>
</tr>
<tr>
<td>RPA</td>
<td>20/10</td>
<td>74</td>
</tr>
<tr>
<td>LV</td>
<td>75/2-11</td>
<td>—</td>
</tr>
<tr>
<td>Ao</td>
<td>78/52</td>
<td>—</td>
</tr>
<tr>
<td>FA</td>
<td>83/52</td>
<td>95</td>
</tr>
</tbody>
</table>

Body weight 62.5 kg
Surface area 1.67 m²
O₂ consumption 196 ml/min
Cardiac index 3.61/min/m²
Pulmonary index 3.61/min/m²
Systemic resistance 22.5 units·m²
Pulmonary resistance 4.2 units·m²
Left-to-right shunt 0% by saturation data <5.0% from dye curves

Abbreviations: SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; RV = right ventricle; PA = pulmonary artery; LPA = left pulmonary artery; RPA = right pulmonary artery; LV = left ventricle; Ao = aorta; FA = femoral artery.

Figure 1. Posteroanterior chest film revealing normal cardiac size and no increase in the pulmonary vascular markings.

Phrenic, lateral thoracic and esophageal arteries as well as from anomalous branches of the aorta. Etiologically, internal mammary artery-to-pulmonary artery fistulas may be classified as congenital or acquired. The acquired variety may be secondary to inflammatory, neoplastic, or traumatic events. A congenital fistula may occur as an isolated event, as in the present case, or in association with another anomaly, as in the case of Árvay et al., with pentology of Fallot. Seven of the 10 previously reported cases were considered to be of congenital origin (table 2).

Figure 2. Simultaneous posteroanterior (A) and lateral (B) frames from a selective left subclavian and internal mammary artery angiogram. (A) Contrast injected into left subclavian artery fills left internal mammary artery (IMA), which becomes tortuous distally and empties into the main pulmonary artery (MPA) just above valve level. Note outline of valve cusps. (B) Lateral view of the same events as in panel A.
An anatomic basis exists for the potential formation of significant connections between the internal mammary artery circulation and the pulmonary circulation through accessory bronchial arteries arising from the former. Anatomic connections between bronchial arteries and pulmonary arteries have been shown even in normal lungs.\(^6\) Congenital fistulas probably develop when the common embryologic anlage of the veins and arteries fails to differentiate properly.\(^1\)\(^,\)\(^4\)

The diagnosis of internal mammary artery-to-pulmonary artery fistula is most often made in young adulthood, with the average age in the reported cases being 26 years. The youngest patient was a 9-year-old child with a clearly congenital fistula,\(^11\) and the oldest patient was a 57-year-old man with a fistula of probably traumatic and inflammatory etiology.\(^9\) The male-to-female ratio is approximately 2:1 (seven males and three females) in the previously reported cases.

Patients are usually asymptomatic upon presentation to the physician but rarely may have symptoms of an associated condition.\(^5\)\(^,\)\(^10\) A continuous murmur over the precordium has been present in all cases and is the basis on which further studies are carried out. The patients are often suspected of having a patent ductus arteriosus or a systemic arteriovenous fistula.\(^5\) Conditions causing continuous precordial murmurs are numerous (table 3), but patent ductus arteriosus is the prototypical one.\(^12\) Other systemic artery-to-pulmonary artery shunts are rare.\(^12\)

### Table 2. Reported Cases of Internal Mammary Artery-to-Pulmonary Artery Fistulas

<table>
<thead>
<tr>
<th>Source</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Signs</th>
<th>Diagnostic procedures</th>
<th>Type and location</th>
<th>Etiology</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Burchell et al. (2017)</td>
<td>20</td>
<td>M</td>
<td>Dyspnea, giddiness</td>
<td>Cyanosis, clubbing, bruit below right nipple</td>
<td>Chest x-ray</td>
<td>Pulmonary AV fistula with connection between right IMA and right PA</td>
<td>Congenital/traumatic</td>
<td>Right middle lobectomy, ligation of vessels</td>
</tr>
<tr>
<td>Voll et al. (1964)(^1)</td>
<td>32</td>
<td>F</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Right-heart cath, selective angio</td>
<td>Right IMA to right PA</td>
<td>Congenital</td>
<td>Partial right middle lobectomy, ligation of vessels</td>
</tr>
<tr>
<td>Árvay et al. (1965)(^1)</td>
<td>9</td>
<td>F</td>
<td>None</td>
<td>Cyanosis, continuous murmur</td>
<td>Selective angio</td>
<td>Right IMA to right middle lobe PA and MPA</td>
<td>Congenital</td>
<td>Unknown</td>
</tr>
<tr>
<td>Kiphart et al. (1967)(^1)</td>
<td>23</td>
<td>M</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Right-heart cath, selective angio</td>
<td>Right IMA to right upper lobe PA</td>
<td>Congenital</td>
<td>Wedge resection of right upper lobe, ligation of vessels</td>
</tr>
<tr>
<td>Stafford et al. (1969)(^4)</td>
<td>21</td>
<td>F</td>
<td>Fever, malaise, headache, chest pain</td>
<td>Continuous murmur</td>
<td>Right-heart cath, pulmonary angio, selective angio</td>
<td>Left IMA to left PA</td>
<td>Congenital</td>
<td>Resection of tip of lingula, ligation of vessels</td>
</tr>
<tr>
<td>Brundage et al. (1972)(^5)</td>
<td>23</td>
<td>M</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Right-heart cath, selective angio</td>
<td>Right IMA to right lower lobe PA</td>
<td>Congenital</td>
<td>Right lower lobectomy, ligation of vessels</td>
</tr>
<tr>
<td>Dunning et al. (1973)(^6)</td>
<td>29</td>
<td>M</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Right-heart cath, selective angio</td>
<td>Right IMA to right PA</td>
<td>Traumatic</td>
<td>Ligation of vessels</td>
</tr>
<tr>
<td>Dunn and Wexler (1974)(^10)</td>
<td>36</td>
<td>M</td>
<td>Flu-like illness</td>
<td>Continuous murmur</td>
<td>Selective angio</td>
<td>Left IMA to left upper lobe PA</td>
<td>Neoplastic</td>
<td>Resection of left upper lobe</td>
</tr>
<tr>
<td>Saito et al. (1975)(^9)</td>
<td>57</td>
<td>M</td>
<td>Engorged chest wall vessels</td>
<td>Continuous murmur</td>
<td>Right-heart cath, aortogram</td>
<td>Right IMA and chest wall vessels to right PA</td>
<td>Traumatic/inflammatory</td>
<td>None</td>
</tr>
<tr>
<td>Laffey et al. (1976)(^11)</td>
<td>19</td>
<td>M</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Pulmonary angio, selective angio</td>
<td>Right IMA to right PA</td>
<td>Congenital</td>
<td>None</td>
</tr>
<tr>
<td>Present case (1979)</td>
<td>18</td>
<td>F</td>
<td>None</td>
<td>Continuous murmur</td>
<td>Right-heart cath, selective angio</td>
<td>Left IMA to main PA</td>
<td>Congenital</td>
<td>None</td>
</tr>
</tbody>
</table>

**Abbreviations:** IMA = internal mammary artery; PA = pulmonary artery; AV = arteriovenous; MPA = main pulmonary artery; cath = catheterization; angio = angiography.

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The major causes for continuous precordial murmurs (table 3) can, in large part, be placed in four categories: (1) aortopulmonary connections, (2) arteriovenous connections, (3) disturbances of the flow pattern in veins, and (4) disturbances of the flow pattern in arteries. A miscellaneous group contains rarer causes of this murmur.

The physical examination of patients with internal mammary artery-to-pulmonary artery fistulas most often reveals only the continuous murmur. An atypical location of the murmur, other than in the second and third left intercostal space, should prompt one to search for causes other than a patent ductus arteriosus.

The cyanosis, clubbing of the digits and polycythemia of classic pulmonary arteriovenous fistula are not present in isolated systemic-to-pulmonary artery shunt. In this series, it was present only in the case of Burchell and Clagett.

The plain chest film is often abnormal, showing increased pulmonary vascular markings or an infiltrate. In four of the 10 reported cases, the radiograph was unremarkable. Electrocardiography does not appear to be helpful in establishing a diagnosis. The procedure of choice for confirming diagnosis and location is selective angiography of the internal mammary vessels. The detection of a left-to-right shunt by means of dye curves, hydrogen appearance times, or oximetry may be unrewarding. Pulmonary angiography may show early washout of the contrast from involved pulmonary artery segments. The right lung is involved more often than the left; however, as in our case and in the case of Arvay et al., the connection may be to the main pulmonary artery with no lung involvement.

The question of surgery for this group of patients remains unanswered. Some authors have recommended that all such patients have definitive surgery because of the risk of rupture, endarteritis, congestive heart failure, and future enlargement of the shunt. It has been pointed out, however, that the young age of the patients, the uncertainty of the long-term efficacy of surgery, and the lack of data on the long-term effects without surgery would suggest a conservative approach. Eight of the 10 patients reported in the literature were operated upon with either ligation alone or resection of lung tissue as well as ligation. The potential hazards of this surgery are pointed out by Burchell and Clagett and Prutzman and Flick. No long-term follow-up on this group of patients is available. It seems most reasonable to recommend surgery on an individual basis, based on the findings in each case. In the patients with neoplasia, complex congenital heart disease and large shunts, surgery seems to be clearly indicated. Perhaps medical follow-up of our patient, who was unoperated, may help provide some answers.

### Acknowledgment

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