Approximately 85% of the patients with tetralogy of Fallot associated with pulmonary atresia have anomalies of the pulmonary vascular tree. The pulmonary arteries are usually of a small caliber, often nonconfluent, and not connected to the right ventricle. In many cases, the proximal branches are stenotic, hypoplastic, or totally absent. The aortopulmonary collateral arteries are frequently present and may supply partially or almost totally the pulmonary blood flow.

The treatment aims at establishing the confluence, normalizing the caliber and distribution of the pulmonary arteries, which may be obtained through the use of shunts and unifocalization of those vessels. Several procedures are usually required before the definitive correction. Dilation with a balloon catheter and use of stents in the pulmonary arteries may represent an important factor for the success of treatment. The operative risk ranges from 0 to 20% depending on the number of shunts and thoracotomies before the definitive correction, which is only possible in 60 to 70% of patients. Recently, some researchers have adopted a radical approach with complete pulmonary arterial reconstruction, closure of the interventricular communication, and placement of a tube in the right ventricular outflow tract. Although the operative risk has been described as small, this approach requires more complex surgical techniques and experience of the surgical team, which are available only in certain centers.

This study aimed at demonstrating the importance of the angiographic study of the pulmonary blood supply in selecting patients with tetralogy of Fallot and pulmonary atresia for total or partial correction of that malformation.

Objectives

To identify the types of pulmonary vascular blood supply in tetralogy of Fallot with pulmonary atresia by the use of hemodynamic studies.

Methods

Fifty-six patients with tetralogy of Fallot and pulmonary atresia, and ages ranging from 20 days to 4 years, underwent cineangiographic study with contrast medium injections in the following vascular structures: 1) wedged pulmonary vein; 2) aortopulmonary collaterals; 3) thoracic aorta; and 4) ductus arteriosus or systemic-pulmonary shunt.

Results

In the 56 patients studied, pulmonary blood was supplied as follows: in 15, by aortopulmonary collaterals; in 36, only by the ductus arteriosus; and in 5, by the ductus arteriosus and aortopulmonary collaterals. The patients were classified into 6 types depending on the type of pulmonary vascular perfusion and the presence or absence of vascular structures that compose the pulmonary circulation in tetralogy of Fallot with pulmonary atresia.

Conclusion

This type of approach enables the obtainment of information necessary for the correct clinicosurgical management of patients, due to the great complexity and extreme variability of the pulmonary blood supply in tetralogy of Fallot with pulmonary atresia.

Key words

pulmonary atresia, tetralogy of Fallot, aortopulmonary collaterals, angiography
The different types of pulmonary blood perfusion were established based on the presence or absence of the following structures: 1) pulmonary trunk; 2) ductus arteriosus; 3) right and left pulmonary arteries and their confluence; and 4) aortopulmonary collaterals.

The angiographic study of the pulmonary blood supply aimed at clarifying the following parameters: 1) to identify the way blood reaches the pulmonary circulation from the systemic circulation; 2) to confirm the presence of the pulmonary trunk; 3) to demonstrate the presence of confluence of the right and left pulmonary arteries; 4) to identify the existence of aortopulmonary collaterals and their connection with the pulmonary arteries; 5) to demonstrate the presence of obstruction in the pulmonary arteries and aortopulmonary collaterals; and 6) to infer the diameter of the pulmonary arteries.

Results

Of the 56 patients studied, 15 had pulmonary blood supplied by aortopulmonary collaterals with the following characteristics: in 2 patients, no central pulmonary arteries were identified and, in 13, they were confluent; in 10 patients, the pulmonary arteries had minuscule dimensions; and, in 3, the caliber was “adequate”. All the remaining 41 patients had pulmonary arteries of “adequate” caliber, whose characteristics were as follows: 36 were confluent and pulmonary blood was supplied only through a tortuous, small-caliber ductus arteriosus in the shape of a comma, forming with the descending aorta an acute angle. In the 5 remaining patients, the pulmonary arteries were not confluent. Pulmonary blood was supplied through the ductus arteriosus to one lung (right or left) and through an aortopulmonary collateral to the contralateral lung (chart I). No patient had the pulmonary flow bilaterally provided by the ductus arteriosus.

The pulmonary arteries were defined as “adequate” when their caliber was sufficient to accept a systemic-pulmonary shunt. Although arbitrary, that definition is based on the clinical/surgical experience of our unit. Therefore, in our material, a pulmonary artery diameter > 3 mm was considered an adequate caliber.

Because of the complexity and extreme variability of the pulmonary blood supply in this malformation, the investigators, throughout the past decades, have proposed different classifications. In our material, we used a classification based on the presence or absence of the following vascular structures and the pulmonary vascular perfusion found in the tetralogy of Fallot with pulmonary atresia: pulmonary trunk, ductus arteriosus, pulmonary arteries (confluence), and aortopulmonary collaterals (tab. I).

In type I, comprising 25 patients, pulmonary blood flow was supplied exclusively through the ductus arteriosus. All patients had pulmonary trunks, the right and left pulmonary arteries were confluent, the blood supply was unifocal, and no aortopulmonary collaterals were identified (fig. 1). That example depicts vasocostriction of the pulmonary arteries, but all lobes of both lungs were perfused. The ductus arteriosus is already dilated (infusion of prostaglandin E1), and the pulmonary trunk is not atretic.

Type II was represented by 11 patients. The pulmonary trunk was atretic and pulmonary blood flow was also supplied only by the ductus arteriosus. The pulmonary arteries were confluent, had an adequate caliber, and the blood supply was unifocal. No aortopulmonary collaterals were identified in this group (fig. 2). In figure 2, the right and left pulmonary arteries are confluent and perfused all segments of both lungs. Different degrees of stenosis are observed in several segments of the pulmonary arteries.

In type III, represented by 13 patients, pulmonary vascular perfusion is exclusively provided by aortopulmonary collaterals, which Anastomose with the confluent pulmonary arteries. The pulmonary trunk is atretic and pulmonary perfusion is multifocal.
arteries were identified in this group. Pulmonary blood perfusion with the pulmonary arteries.

Type IV was represented by 2 patients. No central pulmonary arteries were identified in this group. Pulmonary blood perfusion is exclusively provided by aortopulmonary collaterals (fig. 4). In the 2 examples depicted in fig. 4, even in the late phase of aortography, the central pulmonary arteries are not visualized, and blood supply is provided from several foci, ie, is multifocal.

In type V, represented by 5 patients, pulmonary vascular perfusion is supplied by the ductus arteriosus and aortopulmonary collaterals. In 2 patients, the right pulmonary artery originates from the ductus arteriosus at the right-hand side of the aortic arch, and the left pulmonary artery originates from one collateral of the descending aorta. In 3 patients, the left pulmonary artery originates from the ductus arteriosus at the left-hand side of the aortic arch, and the right pulmonary artery originates from an aortopulmonary collateral (fig. 5). In figure 5, the pulmonary arteries are not confluent, and stenosis may be seen in the connection of the ductus arteriosus with the pulmonary artery and also in the connection of the collateral with the contralateral pulmonary artery. In this group, pulmonary vascular supply is also multifocal.

Type VI, with perfusion through bilateral ductus arteriosus, was not found in our material, although it has already been reported in the literature in patients with isomeric syndromes.

Of the 56 patients, 20 had their pulmonary blood flow supplied by aortopulmonary collaterals (tab. II). Six patients had one aortopulmonary collateral originating from the descending aorta, 7 had 2 aortopulmonary collaterals, 5 had 3 aortopulmonary collaterals, and 2 had 4 aortopulmonary collaterals. An inverse relation was observed between the total number of aortopulmonary collaterals and the size and existence of central pulmonary arteries.

Of the 43 aortopulmonary collaterals, 39 vessels (90.7% – 95%CI = 76.9% to 97.0%) had a defined narrowing at some point of its trajectory or its origin (tab. II, fig. 3, 4, and 5). The most frequent site of stenosis was the connection between the aortopulmonary collateral and the pulmonary artery. In 4 of 20 patients (20% – 95%CI = 6.6% to 44.3%), no aortopulmonary collaterals with stenosis were identified. The other aortopulmonary collaterals without a clearly defined stenosis were frequently tor-
The incidence of incomplete arterial arborization in our material was 29%, and it was particularly found in situations without any confluence of the central pulmonary arteries. When confluence of the right and left branches existed, it did not reach 4%.

The most frequent site of pulmonary artery stenosis was in the central pulmonary artery (tab. III). In 5 patients, the stenosis was located at the origin of the right pulmonary artery, and, in 8 patients, at the origin of the left pulmonary artery. Stenosis of the peripheral pulmonary artery was evidenced in 5 patients. Four patients had stenosis of the pulmonary artery at the site of the systemic-pulmonary anastomosis (fig. 2C).

In some situations, the central pulmonary arteries are not confluent, but are perfused by systemic vessels with one single head of pressure. In this case, blood supply is also unifocal. Usually, the unifocal pulmonary blood supply has confluence of the central pulmonary arteries, which are supplied by a ductus arteriosus. However, the unifocal pulmonary blood supply may be performed through aortopulmonary collaterals that anastomose to the central pulmonary arteries.

Finally, in even rarer situations of unifocal supply, the pulmonary arteries are supplied by multiple collateral arteries acquired as intercostal vessels.

Multifocal pulmonary blood supply may also occur in the presence of pulmonary arterial confluence (fig. 3C and 3D), in which case the confluent pulmonary arteries are not connected to all intrapulmonary arteries in both lungs. This is the most common form of multifocal pulmonary blood supply. Thus, the confluent pulmonary arteries perfused by one or more aortopulmonary collaterals are connected to a certain region of the lung, while the rest of the pulmonary parenchyma is perfused by another aorto-pulmonary collateral or others that anastomose with the rest of the pulmonary segments. Another rarer possibility of multifocal supply in the presence of confluent pulmonary arteries is when the confluent pulmonary arteries are perfused by the ductus arteriosus and the other pulmonary segments are perfused by aortopulmonary collaterals.

The concept of unifocal and multifocal pulmonary blood supply, as described by Macartney et al., was based on the measurement of total pulmonary resistance relative to the aorta, which is usually elevated. That resistance is consequent to several levels of obstruction of the pulmonary arterial tree (fig. 2B and 2C, and tab. III). From the surgical correction viewpoint, what matters is the relative resistance of the vessel that will be connected to the right ventricle, i.e., the central pulmonary artery. Therefore, when the pulmonary blood supply is unifocal, all intrapulmonary arteries are connected to a single head of pressure, as long as no significant stenosis exists in the central and peripheral branches of the pulmonary artery.

Table II - Stenosis of aortopulmonary collaterals in tetralogy of Fallot with pulmonary atresia

<table>
<thead>
<tr>
<th>N° of patients</th>
<th>N AoP-Co</th>
<th>Total AoP-Co</th>
<th>AoP-Co with stenosis</th>
<th>Pts with stenotic AoP-Co</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>1</td>
<td>6</td>
<td>6 (100%)</td>
<td>6 (100%)</td>
</tr>
<tr>
<td>7</td>
<td>2</td>
<td>14</td>
<td>12 (86%)</td>
<td>5 (71%)</td>
</tr>
<tr>
<td>5</td>
<td>3</td>
<td>15</td>
<td>13 (87%)</td>
<td>3 (60%)</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>8</td>
<td>8 (100%)</td>
<td>2 (100%)</td>
</tr>
<tr>
<td>Total=20</td>
<td>-</td>
<td>43</td>
<td>39 (90%)</td>
<td>16 (80%)</td>
</tr>
</tbody>
</table>

AoP-Co - Aortopulmonary collateral; Pts - patients.

Table III - Stenosis of the pulmonary artery in patients with tetralogy of Fallot and pulmonary atresia

<table>
<thead>
<tr>
<th></th>
<th>Total patients</th>
<th>No-PAS</th>
<th>Central pulmonary artery</th>
<th>Origin in the RPA</th>
<th>Origin in the LPA</th>
<th>Peripheral pulmonary artery</th>
<th>Systemic-pulmonary shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAS - pulmonary artery stenosis; RPA and LPA - right and left pulmonary arteries.</td>
<td>54</td>
<td>32</td>
<td>13</td>
<td>5</td>
<td>8</td>
<td>5</td>
<td>4</td>
</tr>
</tbody>
</table>

Discussion

The capillaries that perfuse the pulmonary acini represent the final common path of the pulmonary vascular blood supply. They connect to an intrapulmonary arterial plexus, which branches inside the bronchopulmonary segments, and the pulmonary blood supply may have one or more systemic sources.

When all intrapulmonary arteries connect to the central pulmonary artery, only one single head of pressure perfuses the entire pulmonary parenchyma of both lungs. This type of blood supply is called unifocal (fig. 1 and 2).

In some situations, the central pulmonary arteries are not confluent, but are perfused by systemic vessels with one single head of pressure. In this case, blood supply is also unifocal. Usually, the unifocal pulmonary blood supply has confluence of the central pulmonary arteries, which are supplied by a ductus arteriosus. However, the unifocal pulmonary blood supply may be performed through aortopulmonary collaterals that anastomose to the central pulmonary arteries.

Finally, in even rarer situations of unifocal supply, the pulmonary arteries are supplied by multiple collateral arteries acquired as intercostal vessels.
When the pulmonary blood supply is multifocal, not all intrapulmonary arteries are connected to a single head of pressure. Because different foci certainly have different heads of pressure, the pulmonary regional blood flow is extremely variable, resulting in hypo- and hyperperfused pulmonary segments.

It is worth emphasizing that the aortopulmonary collaterals are vascular structures different from the bronchial arteries. The aortopulmonary collaterals originate only from one systemic artery and run towards the origin of one intrapulmonary artery in the region of the pulmonary hilum or its vicinity (fig. 3, 4, and 5). They are conducting arteries with no nutritional function in terms of the pulmonary parenchyma. The aortopulmonary collaterals usually originate from the anterior face of the aorta (opposed to the origin of the intercostal arteries) and, more rarely, from the right brachiocephalic trunk, the subclavian artery, or even a coronary artery. They differ from the ductus arteriosus in their histological structure and also because the ductus arteriosus is circumscribed to a certain region of the aortic arch. Even when the ductus arteriosus originates from the right-hand side of the aortic arch, it is somewhat opposite to the origin of the brachiocephalic trunk or the subclavian artery.

Acquired collateral pulmonary circulation may occur in any cyanotic congenital heart disease. A consensus still exists in the literature regarding the difference existing between that type of pulmonary blood supply and the aortopulmonary collaterals. From the pathophysiological viewpoint, the most important differentiation is found in the site of anastomosis with the pulmonary circulation. It may be immediately precapillary in the acquired collateral circulation, or in the hilar region in the case of aortopulmonary collateral circulation. Both provide effective pulmonary vascular blood supply. Because acquired collateral circulation never produces heart failure, it is clear that high resistance to blood flow is present between the aorta and the pulmonary arteries. Studies in animals have reported that that phenomenon results from intimal proliferation in the small caliber bronchial arteries.

The ductus arteriosus is a vascular structure that usually originates from the contralateral side of the aortic arch, close to the bifurcation of the brachiocephalic trunk (fig. 2A, 5A, and 5D). Rarely, the ductus arteriosus originates from one anomalous subclavian artery. In the lack of atrial isomerism, the bilateral ductus arteriosus originate from the contralateral side of the aortic arch, it is somewhat opposite to the origin of the brachiocephalic trunk or the subclavian artery.

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Most aortopulmonary collaterals present as tortuous, large caliber vessels with multiple stenoses in their origin, their trajectory, or at the site of anastomosis with the intrapulmonary arteries (tab. 2) (fig. 3, 4, and 5). They never connect to the intercostal arteries. In their trajectory, they accompany the bronchi but never form a plexus around them. Their morphology is the same from the neonatal period until a more advanced age.

The acquired collateral circulation is rare in the first year of life, and, therefore, it was not identified in our material. However, over the years, it has become more developed, mainly after thoracotomy, when adhesion of the visceral and parietal pleurae allows the development of a centripetal collateral circulation, ie, from the thoracic wall to the lung. In the angiographic study, the acquired circulation appears as innumerous thin vessels originating from any thoracic artery. On the other hand, the bronchial arteries are recognized by their relation with the trachea and main bronchi, and by the way they develop a nutritive plexus with the bronchial walls. Recently, this type of circulation has been valued during total surgical correction, because of the respiratory complications observed in the postoperative period. The latter appear after unilateralization and are related to an ischemic process in the respiratory airways, due to the interruption of the tracheobronchial blood supply during the mobilization of aortopulmonary collaterals.

The central pulmonary arteries are more easily identified when they are confluent, because that is the form in which they usually occur. Their confluence associated with a reduction in or absence of their trunk provides a “seagull” configuration when visualized in the frontal plane (fig. 3B). In our material, they were absent in 2 of the 56 patients (fig. 4). In 22 patients, different degrees of stenoses were observed in their trajectory, and, in 4, they were consequent to systemic-pulmonary shunts (tab. III). When the central pulmonary arteries are not confluent, their identification is often difficult. In most cases, the pulmonary trunk is reduced to a fibrous cord, which is connected to the heart; therefore, during cineangiography, a concomitance of movement exists between the central pulmonary arteries and the heart, while the remaining arteries in the mediastinum move together with the lungs.

In the tetralogy of Fallot with pulmonary atresia, the intrapulmonary arteries have normal configuration and distribution. When no confluence of the central pulmonary arteries exists and the intrapulmonary arteries are connected to the aortopulmonary collateral, they may have a distinct configuration, in the shape of a plexus (fig. 5C). This abnormality may result from a hemodynamic disorder consequent to the way they are connected to the aorta.

When a disorder in connection exists, the intrapulmonary arteries may remain isolated and not fuse with the hilar arteries, as usually occurs. As a result, the pulmonary arterial supply to one lobe, or segment, or even part of a segment, remains completely isolated from the rest of the lung. The segmentary or lobar arteries are connected in their proximal portion either to a central pulmonary artery, or to an aortopulmonary collateral, or both (fig. 3C and 3D). The consequence of that disorder is that the pulmonary blood supply becomes compartmentalized. Therefore, each aortopulmonary collateral perfuses only one segment of the lung. The selective contrast medium injection in the aortopulmonary collateral opacifies only that specific region of the lung.

The combination of pieces of information obtained through selective injection of contrast medium into the aortopulmonary collaterals, into the systemic-pulmonary shunts, and into the central pulmonary arteries will enable the accurate determination of the origin of the pulmonary blood supply of each pulmonary segment, and, more important, of the amount of pulmonary parenchyma connected to a certain pulmonary artery. This will reveal whether duplication of the pulmonary blood supply exists, or not, to the same lobe or segment, ie, whether a determined pulmonary region is perfused by more than one source.

Knowing that information is extremely important in the surgical management of those patients. For example, suppose that the right upper lobe is connected to an aortopulmonary collateral, and also to a central pulmonary artery, while the right lower lobe...
is connected only to an aortopulmonary collateral artery. In that situation, one shunt to the right central pulmonary artery or even a duct of a corrective surgery will only increase the pulmonary blood supply to the right upper lobe (fig. 3C and 3D). However, if a communication exists between the perfusion of both lobes, then the surgical procedure will supply blood to the entire right lung.

The prevalence of incomplete pulmonary arterization was high in our patients, mainly when no confluence existed between the right and left pulmonary arteries. On the contrary, incomplete arterization did not reach 5% when confluence of the right branch with the left one existed 31.

Another point identified in our material was that, when confluence of the right and left pulmonary arteries existed, those vessels had a greater caliber. This finding, however, was not confirmed when association of the aortopulmonary collaterals and the central pulmonary arteries existed (chart 1). In that situation, we identified 10 patients with confluent, but "minuscule", pulmonary arteries and innumerable aortopulmonary collaterals (fig. 3A and 3B). The dimensions of the aortopulmonary collaterals were greater in patients without confluence of the right and left pulmonary arteries, and also when the central pulmonary arteries did not exist.

Finally, the total number of aortopulmonary collaterals is inversely related to the size and existence of central pulmonary arteries (fig. 4C and 4D), which had already been demonstrated by Shimazaki et al. 31.

Some of our observations derived from inferences, mainly because of the limitation of the study imposed by our material, which was composed, by more than 50%, of newborn and infant patients. Therefore, the selective contrast medium injection in each collateral in isolation could not be performed, not only due to the excessive amount of contrast medium required, but also due to the excessive prolongation of the procedure in a group of critically ill patients. Therefore, we decided to accept those observations, because a perfect study could hardly be carried out in all patients.

References