A New Surgical Approach to Scimitar Syndrome

Ignacio Lugones, MD, and Rubén García, MD

Fundación Favaloro University Hospital, Buenos Aires, Argentina

A wide variety of surgical strategies has been described to treat scimitar syndrome. However, the incidence of pathway obstruction remains high. An alternative approach is described, in which the pulmonary venous return is widely connected to the left atrium through a tunnel constructed with the in-situ pericardium.


Scimitar syndrome is a rare congenital anomaly characterized by partial or total anomalous pulmonary venous drainage of the right lung to the inferior vena cava (IVC). It can be associated with a number of other abnormalities such as hypoplastic right lung, dextroposition of the heart, atrial septal defect, and anomalous systemic arterial supply to the right lung with or without sequestration. The aim of surgical treatment is to connect the right pulmonary venous return to the left atrium (LA). We describe a new strategy successfully performed in a young adult patient.

Technique

Selective bronchial intubation with a double-lumen endotracheal tube is performed to deflate the right lung during surgery. After median sternotomy, the right pleural space is opened widely. The pulmonary venous drainage is identified. In this case, there was a very short distance between the connection of the lobar branches to the scimitar vein (SV) and its entrance into the IVC in the abdominal cavity. After careful dissection of the pulmonary veins, the pericardium is opened and the right pulmonary artery is cross-clamped. The SV is transected at the level of the diaphragm and both ends are closed with running sutures. Its left lateral aspect is longitudinally opened as much as possible. A large incision is made in the right lateral wall of the pericardium, anterior or posterior to the phrenic nerve. A wide anastomosis is performed between both incisions (Fig 1). Tension and kinking are avoided because the pericardium is mobilized well enough to reach the SV. Aortic and bicaval cannulation is performed and cardiopulmonary bypass is established. At moderate hypothermia, the aorta is cross-clamped and cold cardioplegic solution is delivered to the aortic root. A large vertical left atriotomy is performed. Its left margin is sutured to the posterior pericardium with fine 6-0 polypropylene running suture (Fig 2). From both vertices of the incision, the running suture is continued, attaching the pericardium to the right atrial wall (Fig 3). Superficial bites must be taken in the inner surface of the pericardium to avoid injury to the phrenic nerve. Air is removed just before the suture is completed (Fig 4). The aortic clamp is removed, and the patient is weaned from cardiopulmonary bypass.

The intraoperative transesophageal echocardiogram performed on our patient showed unobstructed pulmonary venous return, with normal low-velocity blood flow.
throughout the pathway. Extubation was achieved in the operating room. The patient recovered uneventfully, without bleeding, arrhythmias, or any other complication, and hospital discharge was possible on the third postoperative day.

The computed angiotomography performed just before hospital discharge confirmed the echocardiographic findings. The three-dimensional reconstruction showed a wide elliptic connection between the LA and the pericardial sac, with a vertical diameter twice as large as the anteroposterior one (Fig 5).

Comment
A variety of vascular reconstructive procedures have been proposed to reroute the pulmonary venous drainage to the LA in scimitar syndrome. Current surgical approaches include different types of baffling [1] and reimplantation [2], with or without graft interposition [3]. The main complication associated with these procedures is pathway obstruction [4]. Factors contributing to this are long and tight tunnels, narrow anastomosis, turbulent blood flow, synthetic grafts, and kinking or distortion of the reconstructed pathway. Therefore, a wide rerouting made of autologous tissue becomes highly desirable. Our technique leaves the pulmonary veins in the posterior pericardium draining directly into the LA through a large anastomosis, thus determining a low risk of stenosis. Moreover, kinking and traction are avoided even in complex anatomies because the pericardium is mobilized to reach the SV in its native position. Damage to the conduction tissue or obstruction of the IVC are also unlikely, and there is no need to open the interatrial septum or the diaphragm. If this operation is performed in young infants or children, the pathway would probably remain patent owing to its large size, despite body growth.

From the technical point of view, it is a straightforward procedure that can be applied in all anatomic variants without the use of synthetic grafts. The IVC cannulation is performed in the usual way and circulatory arrest is not needed. A short period of aortic cross-clamping is used to open the LA and complete the operation.

Special attention must be paid to certain tips and pitfalls. Deep bites in the inner surface of the pericardium must be avoided to prevent damage to the phrenic nerve. Also, bleeding might be harder to control compared with other techniques. Therefore, it is advisable to be very meticulous when performing the running sutures, especially in the LA, as this area will become inaccessible after the correction is completed. Although the results of our
approach are encouraging, long-term follow-up is needed to assess the efficacy of this new technique.

The authors would like to thank Sebastián Viña for his invaluable collaboration with the drawings.

References