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# Surgery for Pulmonary Venous Obstruction After Repair of Total Anomalous Pulmonary Venous Return

François Lacour-Gayet

The major complication and the main cause of reoperation following surgery for total anomalous pulmonary venous return (TAPVR) is the occurrence of pulmonary venous obstruction. Outcomes after surgical repair of TAPVR have greatly improved in the last 10 years; however, the complex forms of TAPVR, particularly when associated with single ventricle and heterotaxy, still carry a high risk of failure. The pathology of pulmonary venous obstruction following TAPVR surgery is a fibrous intimal hyperplasia associated with some medial hypertrophy. There is an increasing severity in the spectrum of lesions from anastomotic stricture to pulmonary vein ostial stenosis and diffuse pulmonary vein stenosis. For anastomotic lesions, revision of the TAPVR anastomosis by patch enlargement of the left atrial anastomosis provides good results. On the contrary, conventional techniques in cases of pulmonary vein ostial stenosis had very poor results. The sutureless repair technique introduced in 1996 provides better midterm results than any other technique, with freedom from mortality and recurrence improving from 65% to 90%. The sutureless technique is described in detail, with emphasis on the need for resection of the pulmonary vein scar tissue and on the different techniques needed respectively on the right and the left side. In our experience, using an atraumatic technique at the initial repair has greatly decreased the occurrence of late pulmonary venous obstruction.
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The results of surgical repair of total anomalous pulmonary venous return (TAPVR) have greatly improved in the last 10 years, in part due to early diagnosis, emergency operation, and non-traumatic surgical technique. In addition, the use of nitric oxide has allowed prevention and control of postoperative pulmonary hypertensive crises that in the past were responsible for significant mortality and morbidity. The complex forms of TAPVR, particularly when associated with single ventricle and heterotaxy, still carry a high risk of failure. The major complication and the main cause of reoperation in TAPVR surgery is the occurrence of pulmonary venous obstruction (PVO).

Denver Children's Hospital, University of Colorado, Denver, CO.
Address reprint requests to François Lacour-Gayet, MD, Cardio-Thoracic Surgery Department, The Children's Hospital, 1056 East 19th Ave, Denver, CO 80218; e-mail: Lacour-Gayet.Francois@tchden.org

## Pulmonary Venous Obstruction Occurrence

PVO may occur as an isolated lesion, but is most commonly seen following repair of TAPVR. When it occurs immediately after surgery, during the first postoperative days, it is related to a restrictive anastomosis. More commonly, PVO occurs after a free delay varying from 2 to several months, frequently after a large anastomosis repair and uneventful postoperative course.

Complex forms of TAPVR are associated with a significantly higher risk of PVO occurrence. A 20% to 50% incidence of postoperative PVO has been reported after repair of TAPVR associated with single-ventricle and right atrial isomerism.[1-3] Mixed type of drainage, aberrant pulmonary venous drainage, association with scimitar syndrome, presence of genetic syndromes, and weight less than 2.5 kg are all associated with increased occurrence of PVO. If PVO seems to be more frequently seen in infra-

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**Table 1** Growing Severity of PVO After Repair of TAPVR

1. **Anastomotic stenosis between the pulmonary veins confluence and the left atrium without individual ostial stenosis.**
2. **Right pulmonary vein ostial stenosis involving the upper and/or the vein. (A single pulmonary vein stenosis is usually well tolerated.)**
3. **Left pulmonary vein ostial stenosis involving the upper and/or the lower vein.**
4. **Bilateral pulmonary vein stenosis involving two, three, or four individual pulmonary venous ostia.**
5. **Diffuse hypoplasia of one or several pulmonary veins.**
6. **Total atresia of several pulmonary vein ostia.**

cardiac type, however, this incidence does not seem significant. The common denominator of most complex TAPVR is a hypoplasia of the common pulmonary venous trunk. Forms of PVO with native intra-hilar pulmonary vein hypoplasia are rare in TAPVR and are not reported in several TAPVR series.[4-6] This pulmonary venous obstructive disease is a poorly understood form of congenital pulmonary vein hypoplasia and carries a very poor prognosis. Use of steroids has been proposed to reduce the risk of late pulmonary venous stenosis.[7] Recently, the group in Toronto[1] has proposed performance of sutureless repair as the initial procedure in patients with hypoplasia of the common pulmonary vein. The critical element of the technique is to perform the atrial anastomosis directly to the pericardium around the incised common pulmonary vein.

Native congenital PVO accounts for 30% of PVO and may represent a substantially greater therapeutic challenge.[5] PVO following non-TAPVR surgery was recently identified in several series.[1,8] The causal mechanism is not well understood, but may be related to pulmonary vein endothelial trauma during the initial surgery or pericardial adhesions. PVO following radiofrequency ablation to correct atrial fibrillation is an iatrogenic complication of cardiac electrophysiology.

# PVO After Repair of TAPVR: Anatomic Lesions and Diagnosis

The pathology of PVO following TAPVR surgery is a fibrous intimal hyperplasia associated with some medial hypertrophy. Depending on the type of PVO, there is an increasing severity in the spectrum of lesions that are variably associated (Table 1). The diagnosis of pulmonary vein ostial stenosis is based on Doppler echocardiography showing continuous flow with a velocity of over 1.5 millisecond at the site of a pulmonary venous ostium. The acceleration is distant from the vein ostia in the presence of isolated left atrial anastomotic stenosis. Selective pulmonary artery branch angiograms clearly show a delay and a stenosis and/or hypoplasia on the late venous return images. Direct catheterization of the ostia may artificially increase the gradient. Magnetic resonance imaging seems particularly useful for the diagnosis; it is the only imaging modality able to visualize still patent portions of the pulmonary veins in the case of ostial atresia. A pulmonary artery pressure over 50 mm Hg is an indication for surgery.

# PVO After Repair of TAPVR: Techniques for Repair

## Standard Techniques

For patients with isolated anastomotic stenosis, revision of the anastomosis by patch enlargement of the left atrial anastomosis is performed through a trans-septal approach. The anterior aspect of the previous anastomosis between the common pulmonary venous trunk and the left atrial wall is incised. A PTFE patch is used to enlarge the anastomosis. The results are excellent.[7]

Ostial stenosis is frequently associated with this lesion. Conventional repair of individual pulmonary vein ostial stenosis by either endarterectomy scar excision, or patch venoplasty (using either pericardium, living atrial tissue, or polytetrafluoroethylene), have been associated with a risk of recurrence ranging from 60% to 90%.[2,4,5,9,10] Patch enlargement of a left pulmonary venous stenosis using the left appendage tissue is still favored by several centers.

## Sutureless Pericardial Repair

We first published this technique in 1996[11] in a patient operated in April 1995 and published a series of seven patients in 1999.[7] The technique is based on a total resection of the pulmonary vein stenotic scar tissue, creating an opening in the left atrial wall that is left open. The operation needs to maintain intact the acquired adhesions between the posterior left atrial wall, the venae cavae, and the pericardium. Minimal dissection is performed posteriorly. While use of circulatory arrest is simpler, bicaval cannulation with full flow CPB is our preferred technique. The SVC is directly cannulated and snared in high position. The inferior vena cava cannula is introduced in low position and is not snared, which allows draining of most of the inferior vena cava flow return. The left atrium is approached through a trans-septal incision. The stenotic pulmonary venous ostia are identified. They can be reduced to pin holes and are often difficult to identify. The right pulmonary venous stenotic tissue is totally resected from inside the left atrium and also from outside below the Sondergaard sulcus. This resection creates a large opening in the left atrial wall that is left open. The stenotic pulmonary vein scar tissue is excised to the level of the pericardial reflection exposing normal pulmonary vein tissue (Figs 1 and 2). The inter-atrium septum is closed as well as the right atrial wall; the left atrial fenestration with the pericardium being left wide open. The remainder of the technique is different on the right and left side.

For right-sided lesions, the right pericardial wall tissue is anastomosed to the right atrial wall, above the left atrial fenestration, creating a left neo-atrial pouch made by the pericardial sac (that is maintained closed by the adhesions from the previous repair), which contains the open right

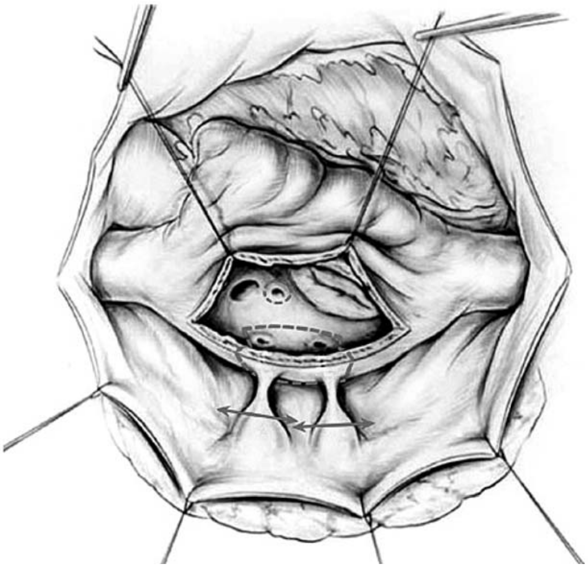

**Figure 1** Resection/sutureless repair (right side): the technique is based on a total resection of the pulmonary vein stenotic scar tissue. Minimal dissection is required posteriorly. The left atrium is approached through a trans-septal incision. The right pulmonary venous stenotic tissue is totally resected. This resection creates a large opening in the left atrial wall that is left open.

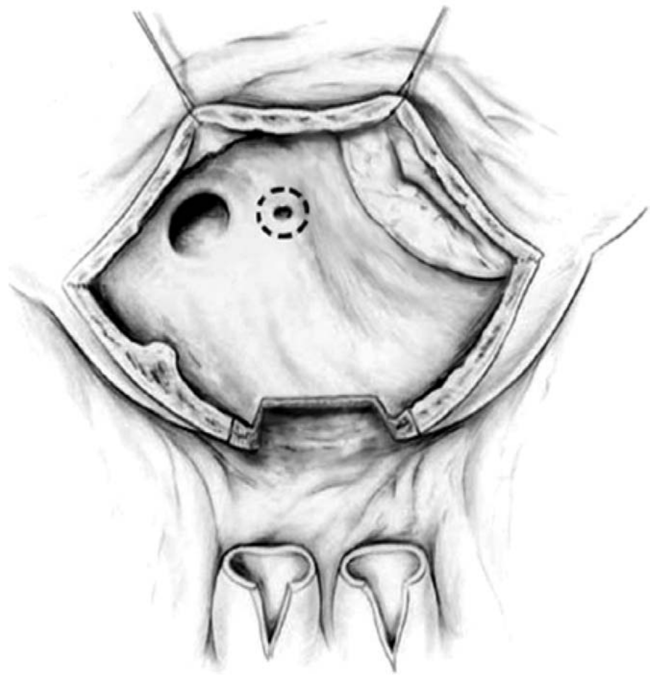

**Figure 2** Resection/sutureless repair (right side): the stenotic pulmonary vein scar tissue is excised to the level of the pericardial reflection exposing normal pulmonary vein tissue.

pulmonary vein draining freely inside the pericardial pouch through the opening of the left atrial wall (Figs 1, 2, and 3). The right phrenic nerve is located below the anastomosis.

For left-sided lesions, the repair is conducted through the left atrial cavity (Figs 4–8). A portion of left atrial tissue is excised around the stenotic scar tissue. This creates a large opening of the left atrial wall. The left pulmonary veins are dissected out to the left pericardium and divided transversely across normal pulmonary vein tissue. In the presence of sufficient pericardial adhesions, no suturing is done (Fig 5). The left pulmonary veins are allowed to drain passively into the left atrium through the posterior pericardial cavity maintained as a closed space by the pericardial adhesions. In the absence of sufficient pericardial adhesions (or in cases with native congenital PVO and intact pericardium), it is necessary to perform an atrio-pericardial anastomosis either from inside the left atrium or from the outside by elevating the heart (division of the inferior vena cava is unnecessary) (Figs 6–8). This suture line is placed on the pericardium, away from the pulmonary venous ostia. The left phrenic nerve is located above the anastomosis.

In 1998, Caldarone and associates[4,9] described a similar technique, leaving in place the pathologic stenotic tissue; the obstructive pulmonary venous lesions being incised longitudinally. Our experience is not in favor of leaving the inflammatory lesion in place. Total resection of all of the scar tissue is more appropriate because the inflammatory stenotic lesion is a process that starts in the left atrium and progresses toward the pulmonary veins. Maintaining the normal pulmonary vein tissue away from the left atrium may reduce the risk of recurrent inflammatory stenosis. Hemorrhage into the pulmonary hilum or inside the pleural cavity has not occurred in our experience with reoperation TAPVR. It has occurred once in a patient with native congenital pulmonary venous stenosis repair with intact pericardium; the hemorrhage was controlled by suturing the pleural tissue to the pericardium.

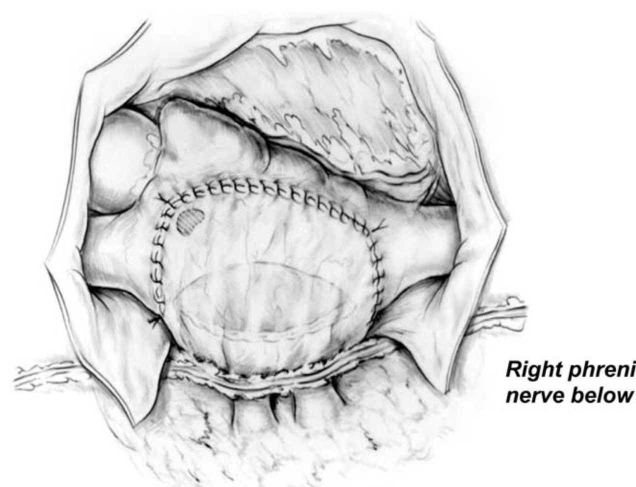


**Figure 3** Resection/sutureless repair (right side): for right-sided lesions, the right pericardial wall tissue is anastomosed to the right atrial wall above the left atrial fenestration, creating a left neo-atrial pouch made by the pericardial sac which contains the open right pulmonary vein draining freely inside the opening of the left atrial wall. The right phrenic nerve is located below the anastomosis.

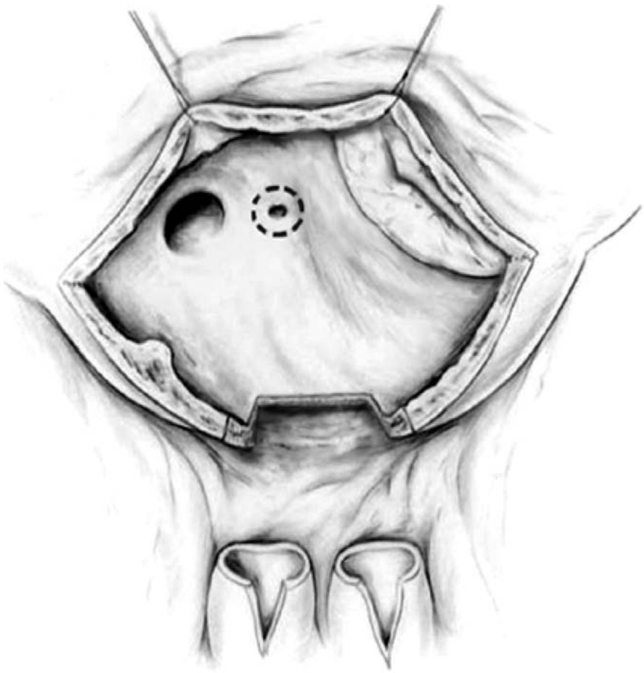

**Figure 4** Resection/sutureless repair (left side, from inside): for left-sided lesions, the repair is conducted through the left atrial cavity. A portion of left atrial tissue is excised around the stenotic scar tissue. This creates a large opening of the left atrial wall.

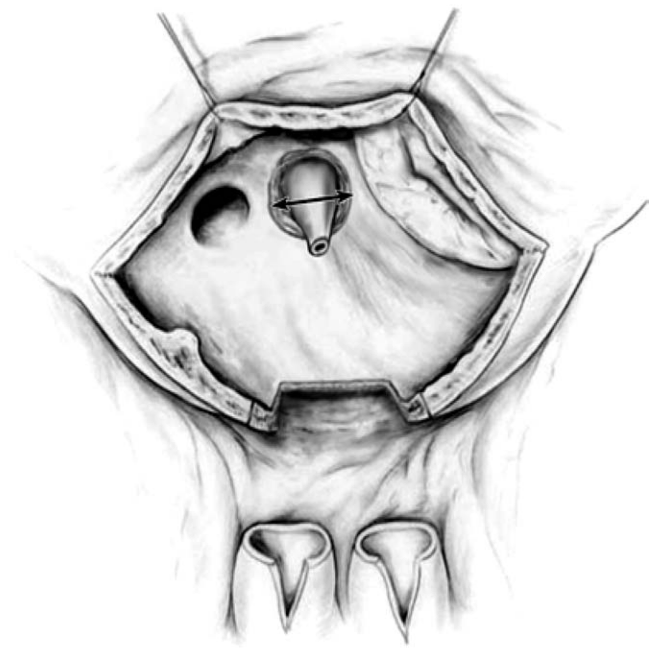

**Figure 5** Resection/sutureless repair (left side, from inside): the left pulmonary veins are dissected out to the left pericardium and divided transversely across normal pulmonary vein tissue. In the presence of sufficient pericardial adhesions, no suturing is done; the left pulmonary veins are allowed to drain passively into the left atrium through the posterior pericardial cavity maintained as a closed space by the pericardial adhesions.

# Outcomes

## Sutureless Pulmonary Vein Repair

The recent literature reports that this original sutureless intra-pericardial pulmonary vein repair provides better midterm results than any other technique[12]; with freedom from mortality and recurrence improving from 65% to 90%.[1,4,5,7] Experience with sutureless repair from the group from Marie Lannelongue in Paris showed satisfactory initial and midterm results.[7] The recent experience for TAPVR repair form the Children's Hospital in Denver is shown on Table 2. There was no mortality in TAPVR with two ventricles and 60% mortality in TAPVR with a single ventricle ($P < .02$). Pulmonary vein obstruction occurred in two patients: an anastomotic stenosis in a supracardiac TAPVR with two ventricles for one patient, and a four-vein ostial stenosis in the other patient with infracardiac TAPVR and single ventricle with heterotaxy occurring 12 months after heart transplantation for failure of the single ventricle. The sutureless repair was performed successfully in two patients: in the patient with a previous heart transplantation (reported at the video session of the World Congress 2005 in Buenos Aires) and in an additional patient referred from outside after two previous repairs of infracardiac TAPVR. The two patients are doing well without recurrence after 30 months and 3 months, respectively.

## Pulmonary Vein Stenting and Cutting Balloon Angioplasty

The results of pulmonary vein stenting have been disappointing. The tendency of the pulmonary veins to produce intimal proliferation makes stenting quite hazardous. Nevertheless, this option remains the last solution in cases of recurrent stenosis after several surgical attempts. Leaving an atrial fenestration to allow angioplasty is preferable. A new generation of devices, such as drug-eluting stents used for coronary artery lesions, may be more effective in the future. Cutting

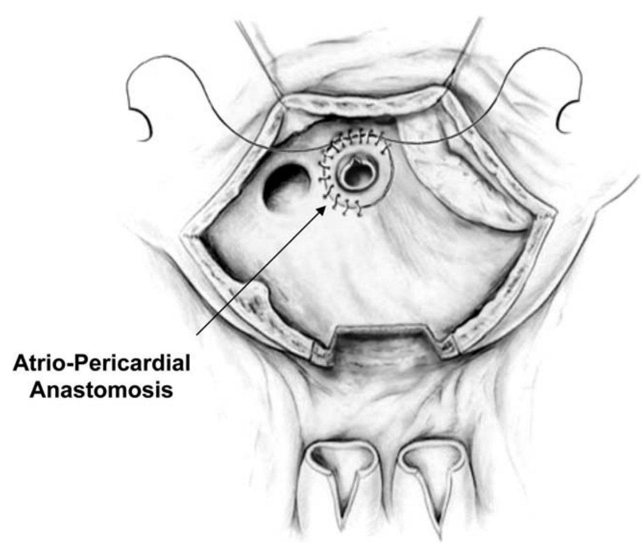


**Figure 6** Resection/sutureless repair (left side/from inside): in the absence of sufficient pericardial adhesions, it is necessary to perform an atrio-pericardial anastomosis from either inside (as shown in this drawing) or outside.

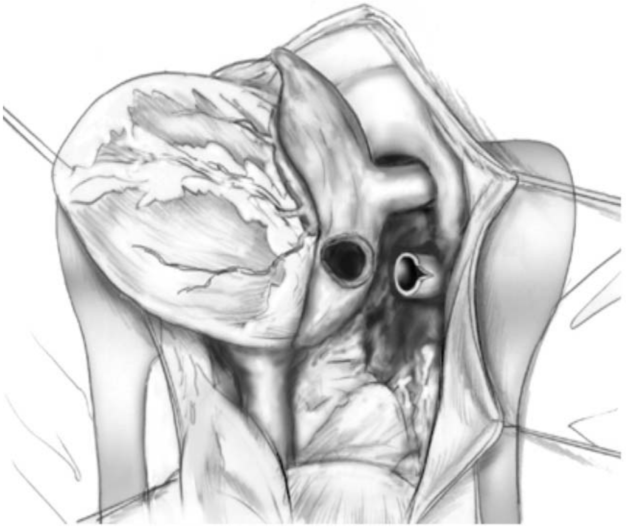

**Figure 7** Resection/sutureless repair (left side/from outside): the atrio-pericardial anastomosis can also be performed from outside by elevating the heart.

**Table 2** Outcome of TAPVR and After Sutureless Intra-Pericardial Repair from the Children's Hospital in Denver

| TAPVR (Total = 23 pts) | Two Ventricles (n = 18) | Single ventricle (n = 5) |
|---|---|---|
| Infra cardiac (n = 11) | 9 | 2 |
| Supra cardiac (n = 6) | 4 | 2 |
| Cardiac (n = 3) | 3 | – |
| Mixed (n = 3) | 2 | 1 |
| SV heterotaxy | – | 4 |
| HLHS + TE fistula | – | 1 |
| Mortality | 0/18 (0%) | 3/5 (60%) $P = .02$ |
| PVO (n = 2) | 1* | 1† |
| Sutureless repair (n = 2) | 1‡ | 1† |
| Mortality (0%) | – | – |

Abbreviations: SV, single ventricle; TE fistula, tracheo-esophageal fistula; PVO, pulmonary vein obstruction.
*Anastomotic stenosis without pulmonary venous ostial stenosis.
†Four pulmonary venous stenosis in TAPVR with single ventricle occurring after heart transplantation.
‡A third patient with infracardiac TAPVR and two ventricles referred from outside was successfully re-operated.

balloon angioplasty has recently been introduced. This technique carries the potential for massive hemorrhage. Furthermore, knowing the tendency of the pulmonary venous endothelium to re-stenose after any manipulation, it is unlikely that the pulmonary veins will stay open more than a few months.

### Prevention of Pulmonary Veins Obstruction Using a "No Touch" Technique

Since we modified our technique of TAPVR repair in 1995 by minimal incision of the pulmonary veins using total circulatory arrest, our rate of PVO has significantly decreased. The neonatal pulmonary venous tissue is clearly very sensitive to inflammatory stenotic lesions for a reason that remains unknown. Our strategy is a "no touch" technique, using circulatory arrest, dissecting minimally the pulmonary vein, and performing an opening incision (strictly limited to the pulmonary venous confluence), and staying well away from the individual pulmonary venous ostia. Another precaution is to use polydioxanone absorbable suture for the anastomosis.

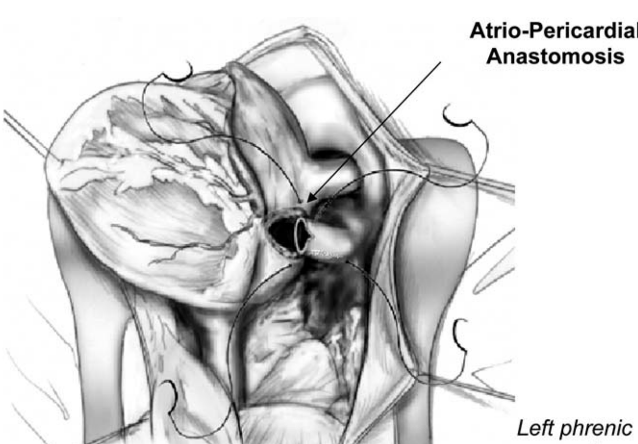


**Figure 8** Resection/sutureless repair (left side, from outside): notice that the suture line is on the pericardium away from the pulmonary vein tissue. The left phrenic nerve is located above.

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