



# **SURGICAL TECHNIQUES FOR PULMONARY ARTERIAL REHABILITATION IN PULMONARY ATRESIA WITH AORTO- PULMONARY COLLATERALS**

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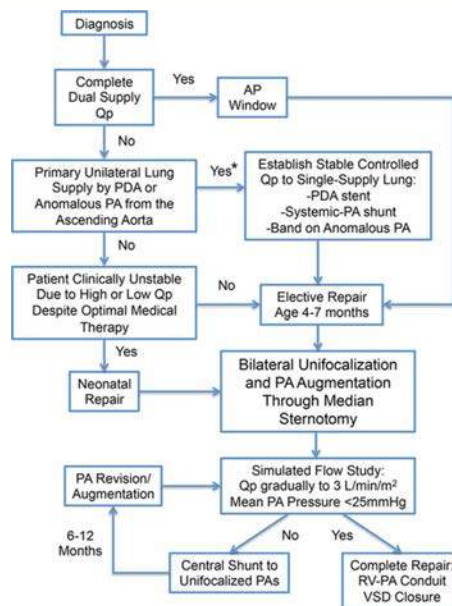
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- I have no relevant disclosures

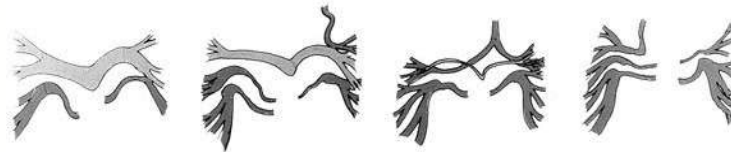


# SOURCE OF PULMONARY BLOOD FLOW

- Pulmonary atresia /Near Pulmonary atresia
  - Ductal dependent supply
  - Major aortopulmonary collateral arteries
  - Ductal dependent and aortopulmonary collateral arteries
  - Antegrade flow/ aortopulmonary collateral arteries

# SPECTRUM OF AORTOPULMONARY COLLATERAL ARTERIAL DISTRIBUTION

- Confluent diminutive central (mediastinal) pulmonary arteries supplying all / near all segments with collateral inflow
- Confluent central (mediastinal) pulmonary arteries supplying some lung segments/ remainder lung segments supplied by aortopulmonary collaterals
- Absence of true central pulmonary arteries/ lung segments supplied by aortopulmonary collaterals



Medium sized

Small

Vestigial  
< 2 mm

Absent

# DESIGNATED FATE OF AORTOPULMONARY COLLATERAL

- Lung segment is only supplied by AP collateral –
  - unifocalization/ incorporation the collateral into the pulmonary circulation
- Lung segment is supplied by AP collateral and native (mediastinal)pulmonary artery
  - Ligate the collateral
  - Unifocalization / incorporation if
    - Stenosis upstream
    - Diminutive connections
    - Addition of native tissue to facilitate growth / reconstruction

# OBJECTIVES OF SURGERY

- Incorporation of blood supply to all segments of the lungs into the antegrade pulmonary circulation by the unifocalization procedure
- Complete repair with closure of the VSD and placement of a conduit from the right ventricle to unifocalized pulmonary arteries
- Goals of achieving normal or near normal pulmonary artery pressures
- Early unifocalization independent of symptoms status

# NEONATAL/ EARLY INFANCY INTERVENTION

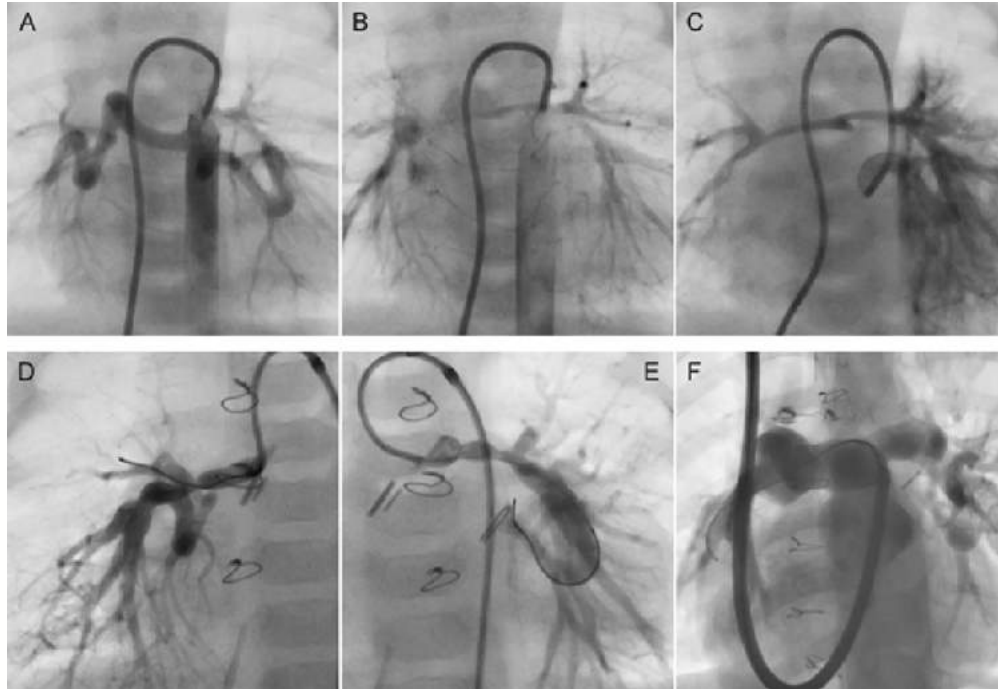
- Central pulmonary with good arborization
- Ductal origin of the left pulmonary artery
- Aortopulmonary collaterals with significant over circulation
- Aortopulmonary collaterals with significant cyanosis



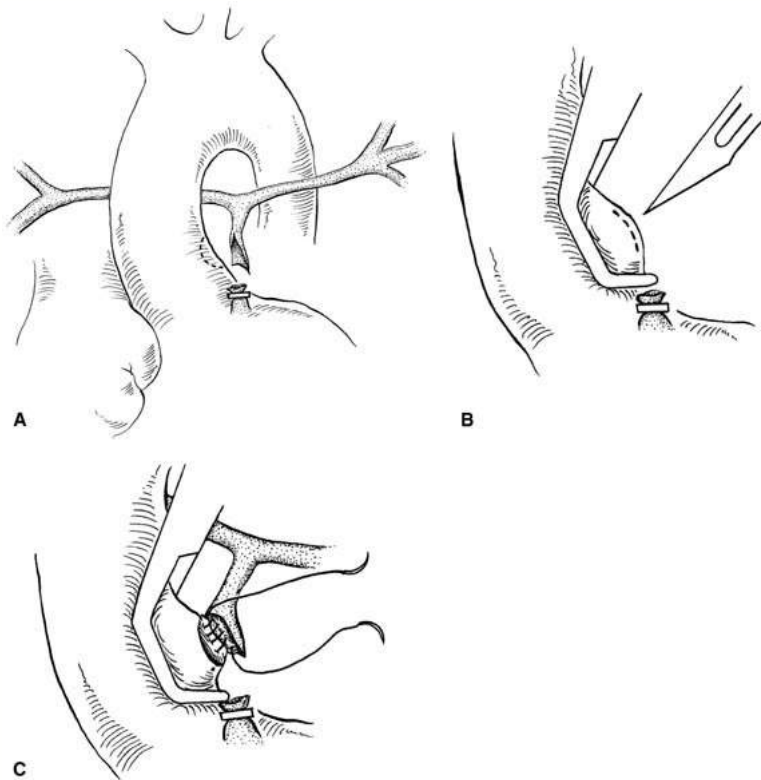
# AORTOPULMONARY WINDOW

- Presence of centrally confluent true pulmonary arteries 1.0 to 2.5 mm in diameter, with well-developed peripheral arborization pattern (central pulmonary arteries all (18) or near all (>15))
- Multiple hypoplastic aortopulmonary collateral vessels, most of which communicated with the true pulmonary arterial system;
- Procedure performed independent of the presence of marked cyanosis.

# AORTOPULMONARY WINDOW



# AORTOPULMONARY WINDOW



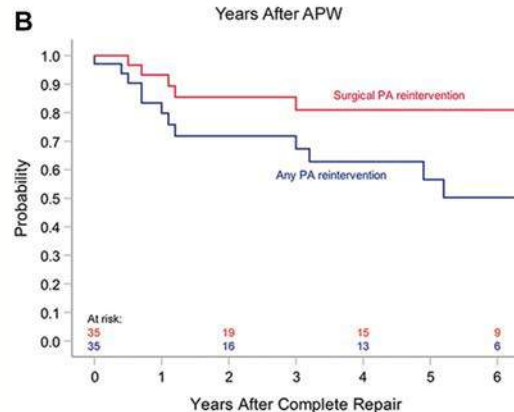
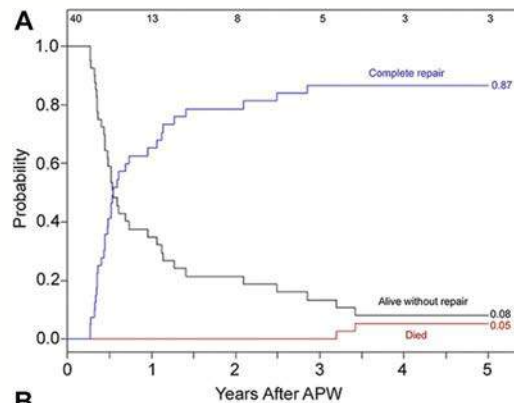
# AORTOPULMONARY WINDOW

- First, the tissue-to-tissue anastomosis greatly reduces the risk of thrombosis.
- Second, the aortopulmonary window is less likely to result in distortion of the branch pulmonary arteries, which by definition are markedly diminutive at the time of this operation.
- Finally, the central location of the aortopulmonary window usually results in balanced blood flow distribution to the right and left branch pulmonary arteries.

# AORTOPULMONARY WINDOW

- Occlude large MAPCAs especially in the setting of relatively high pulmonary blood flow.
- MAPCAs were the sole supply to a portion of lung,
- MAPCAs thought to be dual-supply did not have a robust communication with the PA system,
- Augment the pathway to a particular lung territory if the native PA supplying it did not grow or develop stenosis after APW.

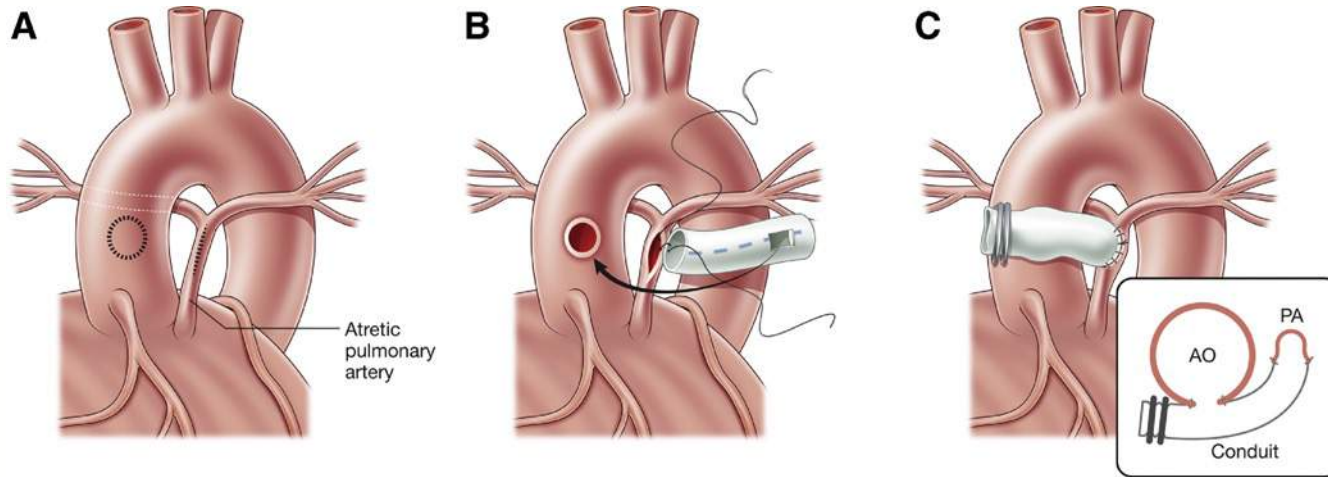
# AORTOPULMONARY WINDOW



# CENTRAL SHUNTS

- Patients who have confluent central branch pulmonary arteries exceeding the size limit of 2.5 mm, (near pulmonary atresia/ vestigial valve remnants present)
- Prosthetic shunt will regulate flow and pressure and lessen the possibility of excessive pulmonary blood flow or a reperfusion injury to the lung, or both.

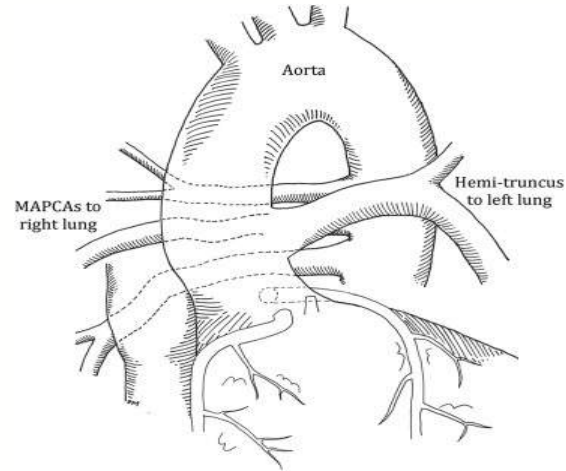
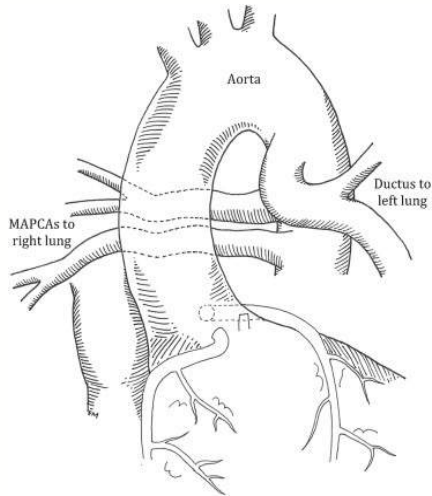
# CENTRAL GORE-TEX SHUNT (LAK'S MODIFICATION)





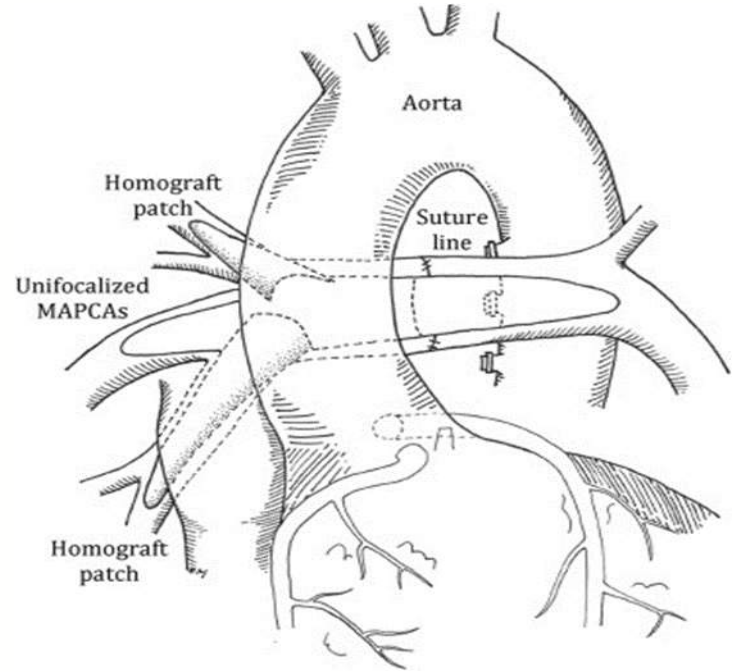
# MAPCAS/ DUCTAL DEPENDENT PULMONARY ARTERY

- Ductal origin to unilateral lung or hemi truncus/ MAPCAS contralateral lung



# MAPCAS/ DUCTAL DEPENDENT PULMONARY ARTERY

- Adequately sized right sided MAPCAs
- Complete repair
- Early Infancy
- Prolonged Hospitalization

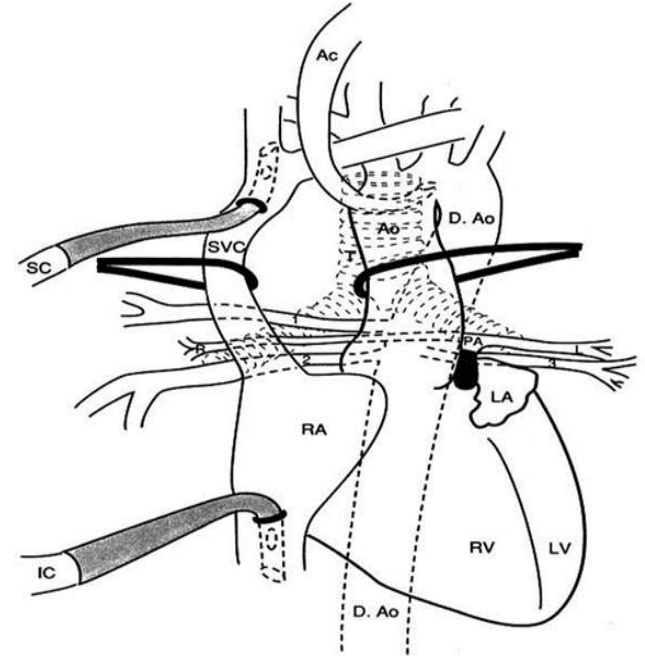


# MAPCAS / DUCTAL DEPENDENT PULMONARY ARTERY

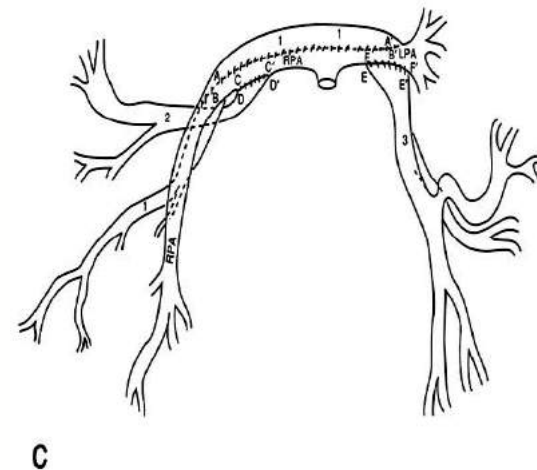
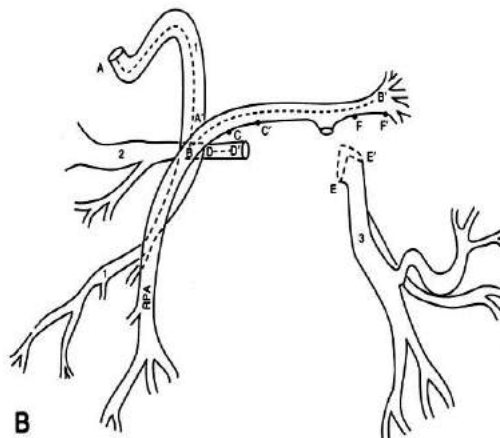
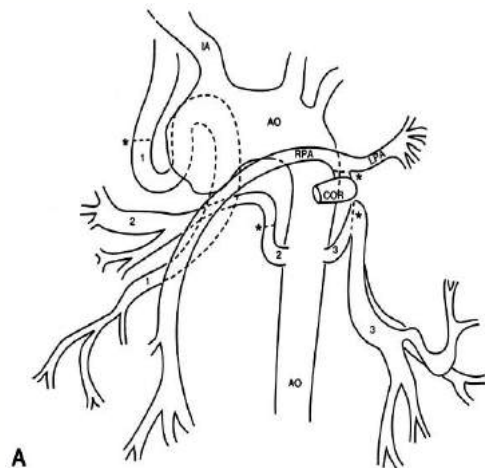
- Small right sided MAPCAs
- Palliative Options – Ductal Stent vs Surgical Shunt to unilateral lung
  - Loss of potential viable proximal pulmonary artery tissue
- Palliative Options- Right sided MAPCAs unifocalization to central shunt

# UNIFOCAIZATION PROCEDURE

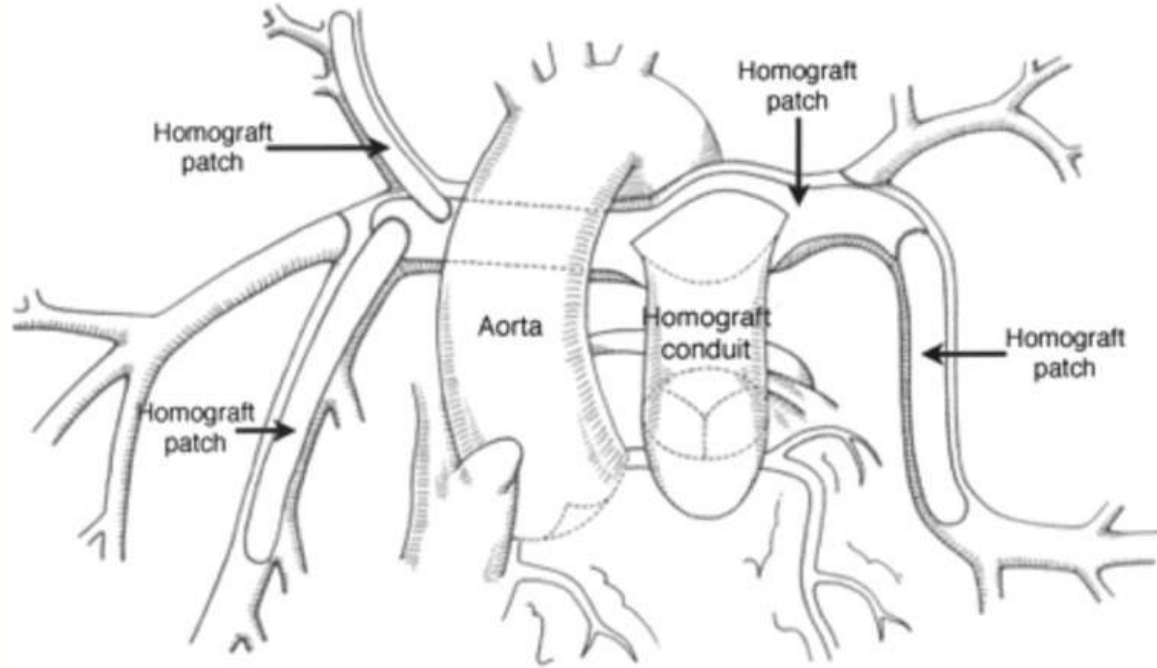
- Transverse sinus approach
- Tracheobronchial angle / dome of left atrium
- Identification/ control of individual MAPCAs
- Mobilization of MAPCAs
- Wide opening of pleura posterior to phrenic



# UNIFOCAIZATION PROCEDURE

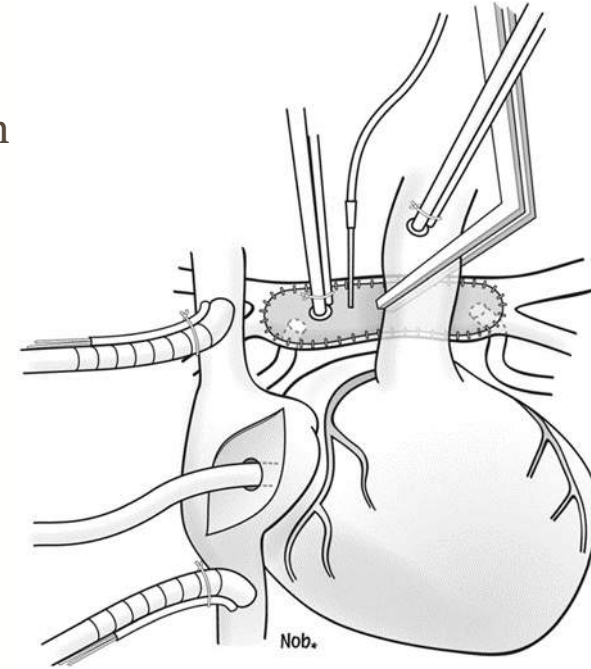


# UNIFOCAIZATION PROCEDURE



# PULMONARY FLOW STUDY

- Assess pulmonary vascular resistance following unifocalization
- 3 liters (2.5)/ min/m<sup>2</sup> to the pulmonary bed
- Acceptable pressure in the PA bed 25mmHg (30)
- Venting of the left atrium / gentle ventilation

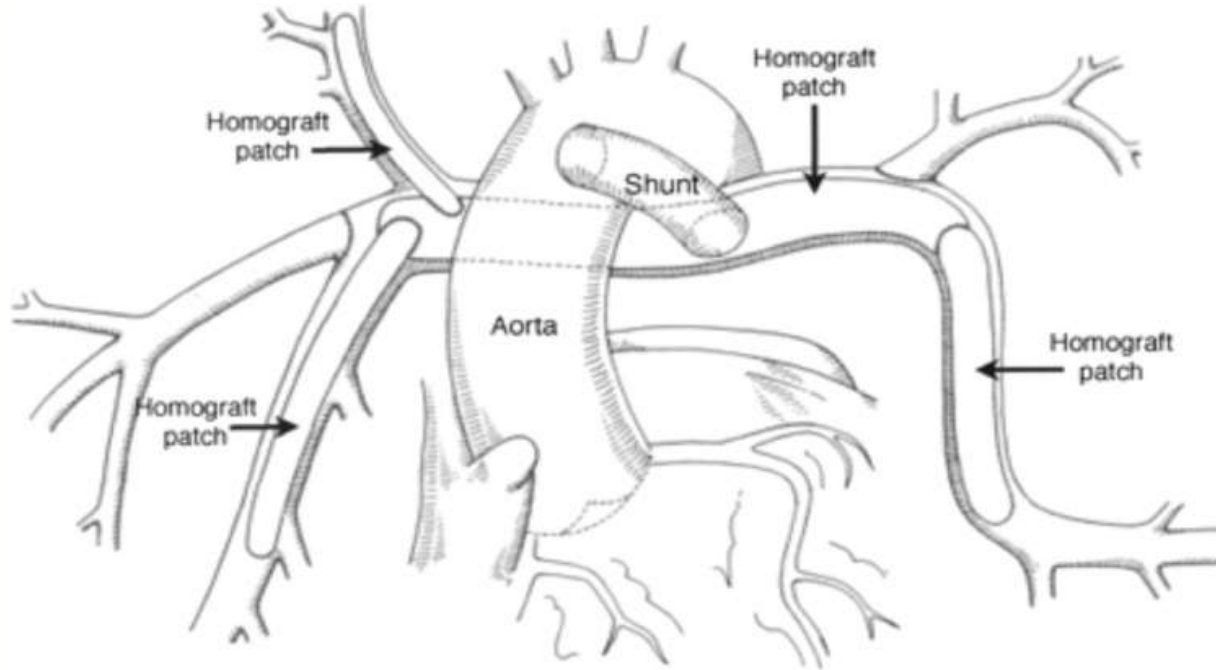


# PULMONARY FLOW STUDY

- Saturations  $>85\%$
- Saturation  $< 82\%$  and older children

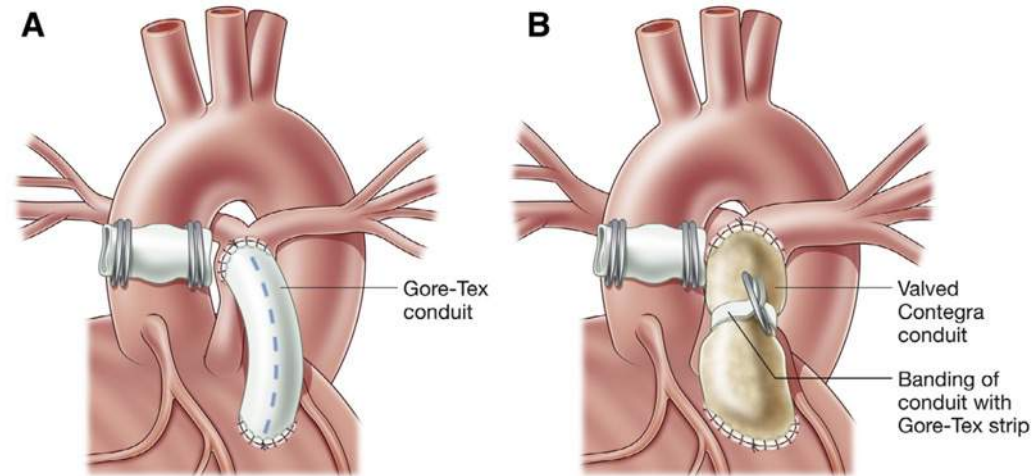


# UNIFOCALIZATION PROCEDURE



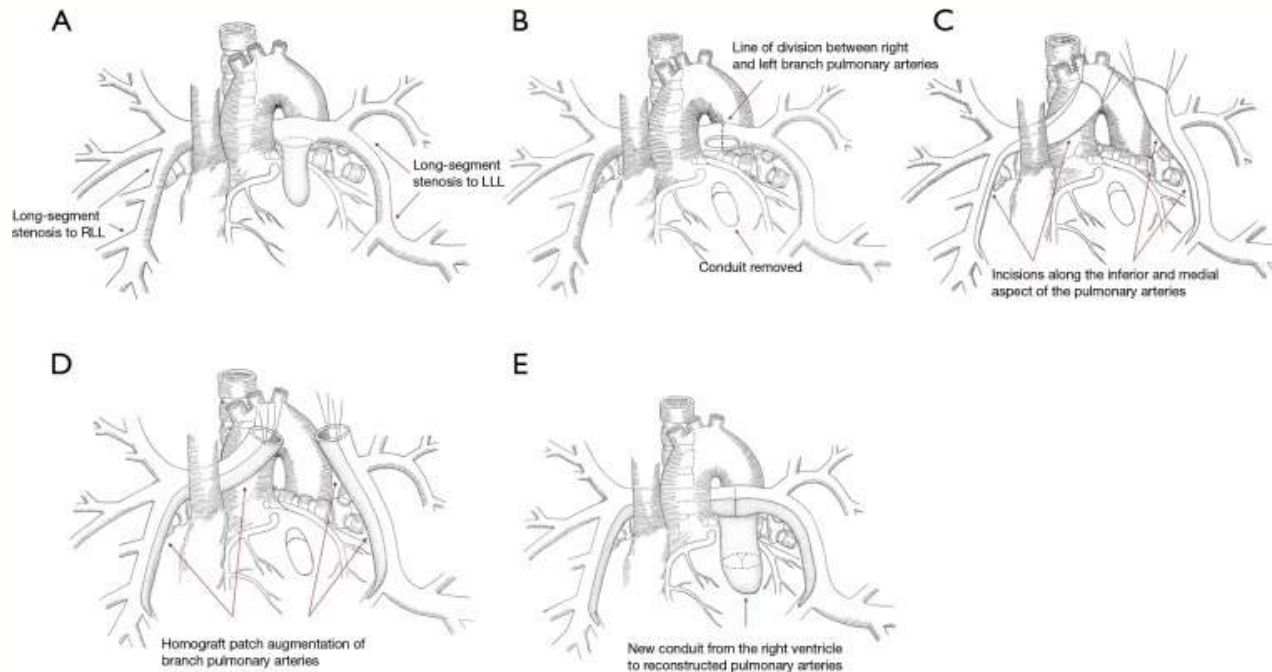
# CENTRAL SHUNT VS RV PA CONDUIT

- Tethering /proximal stenosis
- Uncontrolled pulmonary blood flow
- Right ventricular dysfunction
- Pseudo aneurysm of the RV

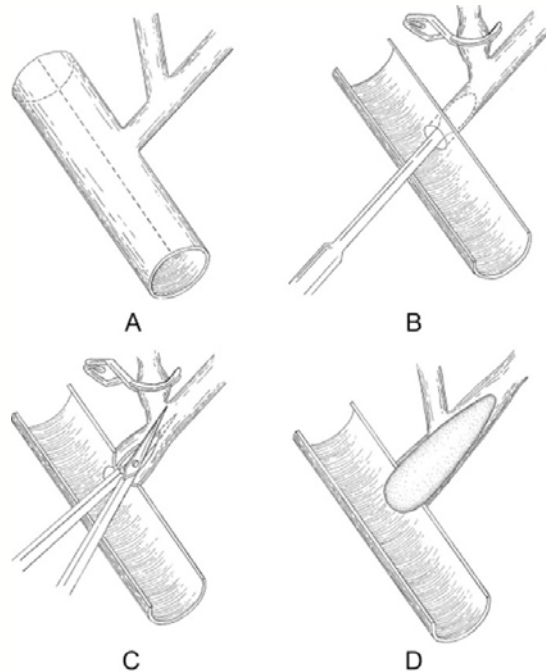
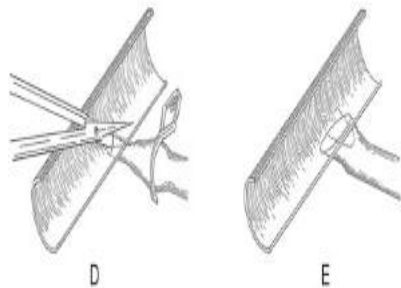
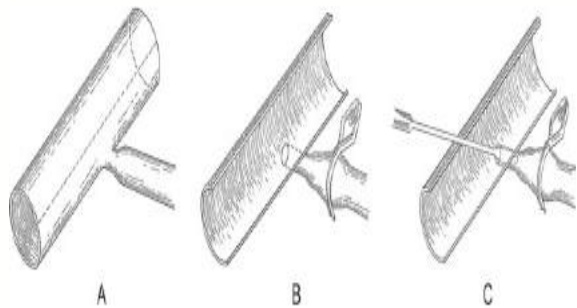


**Figure 4** Right ventricle to pulmonary artery conduit implantation.

# UNIFOCAIZATION REVISION



# SEGMENTAL ARTERY REHABILITATION



# IDEAL STATE

- Complete Repair
- Survival/ Quality of Life
- Normal pulmonary pressures

# Comments/ Thank you