# Pulmonic Valve Disease

Chapter 278 | Part 6: Disorders of the Cardiovascular System

KEY CLINICAL POINTS

1. Pulmonic stenosis (PS) is primarily a congenital disorder; rheumatic involvement is very rare.
2. Severe PS is defined by a peak systolic gradient >64 mmHg (mean >35 mmHg) or Doppler jet velocity >4 m/s.
3. The hallmark of pulmonic regurgitation (PR) is the Graham Steell murmur: a high-pitched, decrescendo diastolic murmur along the left sternal border.
4. Balloon valvuloplasty is indicated for symptomatic patients with moderate or severe PS and for asymptomatic patients with a peak gradient >64 mmHg (mean >35 mmHg).
5. Transcatheter pulmonic valve replacement is an option for severe PR in patients with repaired congenital heart disease (e.g., tetralogy of Fallot).
6. RV dysfunction from afterload mismatch occurs earlier in PS than LV dysfunction in aortic stenosis due to the RV's poor adaptation to pressure overload.
7. Destabilizing triggers (atrial fibrillation, fever, infection, anemia, or pregnancy) can trigger angina or syncope in severe PS.
8. Diuretics are used for right heart failure symptoms provided PR is not moderate to severe.
9. Noonan syndrome (PTPN11 mutation) often involves dysplastic pulmonic valves requiring surgical intervention.
10. In tetralogy of Fallot survivors, a freely regurgitant RV-PA conduit may produce a misleadingly low-pitched murmur despite significant PR.

## 1. DEFINITION & OVERVIEW

- **Pulmonic Valve Disease:** encompasses two primary valvular lesions: pulmonic stenosis (PS) and pulmonic regurgitation (PR).
- **Pulmonic Stenosis (PS):** essentially a congenital disorder.
- **Pulmonic Regurgitation (PR):** may develop as a consequence of primary valve pathology, annular enlargement, or both.
- **Post-Surgical PR:** can occur after surgical treatment of RVOT obstruction in children (e.g., tetralogy of Fallot) or after percutaneous pulmonic balloon valvotomy.
- **Rheumatic Involvement:** The pulmonic valve is only very rarely affected by the rheumatic process.

### 1.1 Classification by Lesion Type

- **Pulmonic Stenosis (PS):** Obstruction of right ventricular outflow.
- **Pulmonic Regurgitation (PR):** Backflow from pulmonary artery to right ventricle during diastole.
- **Mixed Disease:** Carcinoid typically causes mixed pulmonic valve disease with both PR and PS.

### 1.2 Classification by Etiology

- **Congenital:** The most common cause of PS.
- **Acquired:** Carcinoid, tumor, endocarditis, post-valvotomy, annular enlargement, pulmonary hypertension.
- **Syndromic:** Noonan syndrome (dysplastic valves).
- **Idiopathic:** Idiopathic dilation, Marfan syndrome.

---

# 2. ETIOLOGY & PATHOPHYSIOLOGY

- **Hemodynamics of PS:** Defined by a systolic pressure gradient between the right ventricle (RV) and the main pulmonary artery (PA).
- **RV Adaptation:** RV dysfunction from afterload mismatch occurs earlier in PS than LV dysfunction in aortic stenosis (AS) due to the RV's poor adaptation to pressure overload.
- **Right Atrial (RA) Dynamics:**
  - a wave elevates due to higher pressures needed to fill a noncompliant, hypertrophied RV.
  - prominent v wave signifies functional tricuspid regurgitation (TR) from RV and just as in aortic regurgitation (AR), it indicates annular dilation.
- **Regurgitation Pathophysiology:**
  - Severe PR results in RV chamber enlargement and eccentric hypertrophy.
  - PR is a state of increased preload and afterload.
  - The diastolic pressure gradient between the PA and RV decreases throughout diastole, resulting in a decrescendo murmur.
  - As RV diastolic pressure increases, the murmur becomes shorter in duration.
- **Cardiac Output (CO):**
  - Forward CO is preserved during early stages but may not increase with exercise and declines over time.
  - Reduction in RV ejection fraction is an early indicator of hemodynamic compromise.
- **Advanced Disease:** Significant enlargement of RV and RA with marked elevation of jugular venous pressure.
- **Pulmonary Hypertension (PH):** Long-standing severe PA hypertension can result in dilation of the pulmonic valve ring and PR.
- **Incidental Findings:** Trace or mild PR of no hemodynamic or clinical consequence is frequently observed on TTE in the absence of structural pulmonic valve disease.

Table 278-1: Causes of Pulmonic Valve Disease

- **Pulmonic Stenosis:** Congenital, Carcinoid, Tumor, Endocarditis.
- **Pulmonic Regurgitation:** Primary valve disease, Post-valvotomy, Annular enlargement, Pulmonary hypertension, Marfan syndrome, Idiopathic dilation, Carcinoid, Tumor, Endocarditis.

## 2.1 Hemodynamic Definitions

- **Severe PS:** Peak gradient >64 mmHg (mean >35 mmHg) or Doppler jet velocity >4 m/s.
- **Moderate PS:** Peak gradient 36–64 mmHg (Doppler jet velocity 3–4 m/s).
- **Mild PS:** Peak gradient <36 mmHg (Doppler jet velocity <3 m/s).

## 2.2 Pathophysiology of Regurgitation

- **Structural Impact:** Severe PR results in RV chamber enlargement and eccentric hypertrophy.

- **Hemodynamic State:** PR is a state of increased preload and afterload.
- **Murmur Dynamics:** Diastolic pressure gradient between PA and RV decreases throughout diastole → decrescendo murmur.
- **Clinical Progression:** Forward CO preserved early → may not increase with exercise → declines over time.
- **Early Warning:** Reduction in RV ejection fraction is an early indicator of hemodynamic compromise.

### 2.3 Pathophysiology of Stenosis

- **RV Hypertrophy (RVH):** Develops from sustained obstruction; systolic ejection is prolonged.
- **Comparison to AS:** RV dysfunction occurs earlier in PS than LV dysfunction in AS due to poor adaptation to pressure overload.
- **RA Dynamics:**
  - a wave elevation → noncompliant, hypertrophied RV.
  - v wave prominence → functional tricuspid regurgitation (TR).

---

## 3. CLINICAL FEATURES

- **General Presentation:**
  - Mild or moderate PS are usually asymptomatic.
  - Mild or moderate PR do not, by themselves, result in symptoms; other issues (e.g., PA hypertension) may dominate the clinical picture.
- **Severe Disease Symptoms:**
  - **PS:** Exertional dyspnea or early-onset fatigue.
  - **Very Severe PS:** Anginal chest pain and syncope (due to RV oxygen supply-demand mismatch).
  - **PR:** Fatigue, exertional dyspnea, abdominal fullness/bloating, and lower extremity swelling.
- **Right Heart Failure:** Signs such as hepatomegaly, ascites, and edema are uncommon but may appear very late in the disease.

### 3.1 Symptoms of Pulmonic Stenosis

- **Mild/Moderate:** Asymptomatic.
- **Severe:** Exertional dyspnea, early-onset fatigue.
- **Very Severe:** Anginal chest pain, syncope.
- **Destabilizing Triggers:** Atrial fibrillation, fever, infection, anemia, pregnancy.

### 3.2 Symptoms of Pulmonic Regurgitation

- **Mild/Moderate:** Asymptomatic.
- **Severe:** Fatigue, exertional dyspnea, abdominal fullness/bloating, lower extremity swelling.
- **Dominant Features:** Often dominated by symptoms of pulmonary artery hypertension.

### 3.3 Physical Findings in Pulmonic Stenosis

- **Murmur:** Mid-systolic, crescendo-decrescendo; heard best in the left second interspace.
- **Ejection Sound:** Often preceded by an ejection sound (click) in younger adults with pliable valves.
- **Progression:** As severity increases, the ejection sound moves closer to S1 and eventually becomes inaudible.
- **S2 Component:** Pulmonic valve closure is delayed; P2 component is reduced or absent.

- **JVP:** Prominent a wave (noncompliant RV).
- **Palpation:** Parasternal or RV lift may be felt with significant pressure overload.

### 3.4 Physical Findings in Pulmonic Regurgitation

- **Graham Steell Murmur:** High-pitched, decrescendo diastolic murmur along the left sternal border.
  - Note: Can be difficult to distinguish from aortic regurgitation (AR).
  - Observation: May become louder with inspiration.
- **Associated Signs:** Usually associated with a loud/palpable P2 and an RV lift.
- **Tetralogy of Fallot Survivors:**
  - RV-PA conduit may be freely regurgitant (no valve present).
  - Result: Diastolic murmur can be misleadingly low-pitched and short duration despite significant PR and RV volume overload.

---

## 4. INVESTIGATIONS & DIAGNOSIS

1. **Transthoracic Echocardiography (TTE):** Primary tool for definitive diagnosis, characterization of valve, jet velocity, gradient, RV function, and PA pressures (which should be low).
2. **Transesophageal Echocardiography (TEE):** Useful for improved delineation of the RVOT and assessment of infundibular hypertrophy.
3. **Cardiac Magnetic Resonance (CMR):** Provides greater anatomic detail; used for precise assessment of RV volumes and function, especially in patients with repaired congenital heart disease.
4. **Electrocardiography (ECG):** Identifies right axis deviation, RVH, and RA enlargement in adult patients with severe PS.
5. **Chest Radiography:**
   - Frontal Plane: Poststenotic dilation of the main PA.
   - Lateral Film: Filling of the retrosternal airspace due to RV enlargement.
   - Other: Potential elevation of cardiac apex off left hemidiaphragm; RA may be enlarged.
6. **Cardiac Catheterization:**
   - Not usually necessary for diagnostic purposes.
   - If performed (e.g., during planned transcatheter procedure): Measure pressures just below and above the pulmonic valve.
   - Note: Correlation between Doppler peak instantaneous gradient and catheterization peak-to-peak gradient is weak; peak-to-peak may correlate better with the Doppler mean gradient.

### 4.1 Electrocardiography

- **Findings:** Right axis deviation, RVH, RA enlargement.
- **Context:** Seen in adult patients with severe PS.

### 4.2 Chest Radiography

- **Frontal Plane:** Poststenotic dilation of the main PA.
- **Lateral Film:** Filling of the retrosternal airspace due to RV enlargement.
- **Apex:** In some patients with RVH, the cardiac apex appears to be lifted off the left hemidiaphragm.
- **RA:** The RA may also be enlarged.

### 4.3 Echocardiography

- **TTE:** Definitive diagnosis and characterization; assesses valve, jet velocity, gradient, RV function, and PA pressures.
- **TEE:** Useful for improved delineation of the RVOT and assessment of infundibular hypertrophy.

### 4.4 Cardiac Catheterization

- **Necessity:** Not usually necessary for diagnostic purposes; performed as part of planned transcatheter PV procedure.
- **Measurement:** Pressures obtained from just below and above the pulmonic valve.
- **Correlation:** Correlation between Doppler peak instantaneous gradient and catheterization peak-to-peak gradient is weak.
  - Note: Catheterization peak-to-peak gradient may correlate better with the Doppler mean gradient.

### 4.5 Cardiac Magnetic Resonance

- **Utility:** Provides greater anatomic detail.
- **Indication:** Particularly in patients with repaired congenital heart disease.
- **Assessment:** More precise assessment of RV volumes and function.

---

## 5. MANAGEMENT & TREATMENT

1. **Medical Management:**
   - Diuretics used to treat symptoms and signs of right heart failure.
   - Condition: Provided PR is not moderate to severe.
2. **Interventional Management (Pulmonic Stenosis):** Percutaneous pulmonic balloon valvuloplasty.
   - Indications:
   - Symptomatic patients with moderate or severe PS.
   - Asymptomatic patients with a peak gradient >64 mmHg (mean >35 mmHg).
3. **Surgical Management:**
   - Required when the valve is dysplastic (e.g., Noonan syndrome).
4. **Management of Pulmonary Hypertension:**
   - For significant functional PR due to PA hypertension and annular dilation.
   - Strategy: Pursue reduction of pulmonary vascular resistance/pressure via pharmacologic/vasodilator and/or surgical/interventional strategies, depending on the cause of the PA hypertension (e.g., idiopathic PA hypertension, left-sided heart valve disease).
5. **Transcatheter Pulmonic Valve Replacement:**
   - Indicated for severe PR in patients with repaired congenital heart disease (e.g., tetralogy of Fallot, PS, or atresia).
   - Note: Procedure was introduced clinically prior to transcatheter aortic valve replacement; modifications made to address earlier concerns regarding infective endocarditis.

### 5.1 Management of Pulmonic Stenosis

- **Medical:** Diuretics for right heart failure.
- **Interventional:** Percutaneous pulmonic balloon valvuloplasty (Symptomatic moderate/severe OR asymptomatic >64 mmHg).
- **Surgical:** Required for dysplastic valves (e.g., Noonan syndrome).
- **Coordination:** Multidisciplinary heart team for treatment decisions.

### 5.2 Management of Pulmonic Regurgitation

- **Medical:** Diuretics for right heart failure.
- **Pulmonary Hypertension Management:** Reduce pulmonary vascular resistance/pressure (pharmacologic or surgical depending on cause).
- **Transcatheter Replacement:** For severe PR after childhood repair (e.g., tetralogy of Fallot).

---

## 6. PROGNOSIS & COMPLICATIONS

- **Progression of Stenosis:**
  - Mild PS: Rarely progresses.
  - Moderate PS: May worsen with age due to valve thickening and calcification.
- **Complications of Regurgitation:**
  - RV/RA Enlargement: Significant enlargement in advanced stages.
  - JVP: Marked elevation of jugular venous pressure.
  - Cardiac Output: Forward CO declines over time.
  - Early Warning: Reduction in RV ejection fraction indicates hemodynamic compromise.
- **Right Heart Failure:** Symptoms (hepatomegaly, ascites, edema) appear very late in the disease.

### 6.1 Progression of Stenosis

- **Mild PS:** Rarely progresses.
- **Moderate PS:** May worsen with age due to valve thickening and calcification.

### 6.2 Complications of Regurgitation

- **RV Enlargement:** Significant enlargement of RV and RA.
- **JVP:** Marked elevation of jugular venous pressure.
- **CO Decline:** Forward CO declines over time.
- **RV Function:** Reduction in RV ejection fraction is an early indicator of compromise.

---

## 7. SPECIAL CONSIDERATIONS

- **Noonan Syndrome:**
  - Mutation: PTPN11 gene.
  - Clinical Feature: Dysplastic pulmonic valves.
- **Tetralogy of Fallot Survivors:**
  - Anatomy: RV-PA conduit may be freely regurgitant because it lacks a valve.
  - Hemodynamics: PA pressures are not elevated.
  - Murmur: Diastolic murmur can be misleadingly low-pitched and short duration despite significant PR and RV volume overload.
- **Destabilizing Triggers:**
  - Conditions: Atrial fibrillation, fever, infection, anemia, or pregnancy.
  - Effect: Can precipitate angina or syncope in patients with very severe forms of obstruction.

### 7.1 Genetic Syndromes

- **Noonan Syndrome:** PTPN11 mutations; dysplastic pulmonic valves.

### 7.2 Post-Surgical Considerations

- **Tetralogy of Fallot:** RV-PA conduit may be freely regurgitant (no valve) → low-pitched, short duration diastolic murmur despite significant PR and RV volume overload.

### 7.3 Destabilizing Triggers

- **Angina/Syncope Risk:** Very severe obstruction + Trigger (AFib, fever, infection, anemia, pregnancy) → increased risk of angina or syncope.

---

## 8. KEY PEARLS & CLINICAL TRAPS

- **Pulmonic Stenosis:** Primarily a congenital disorder; rheumatic involvement is rare.
- **RV vs LV:** RV dysfunction from afterload mismatch occurs earlier in PS than LV dysfunction in AS due to poor RV adaptation.
- **Graham Steell Murmur:** High-pitched, decrescendo diastolic murmur is the hallmark of PR.
- **Balloon Valvuloplasty:** Indicated for symptomatic moderate/severe PS or asymptomatic patients with peak gradient >64 mmHg (mean >35 mmHg).
- **Transcatheter Replacement:** Option for severe PR in patients with repaired congenital heart disease (e.g., tetralogy of Fallot).
- **Diuretics:** Used for right heart failure symptoms provided PR is not moderate to severe.
- **Destabilizing Triggers:** Pregnancy, fever, infection, anemia, or atrial fibrillation can trigger complications in severe PS.
- **Tetralogy of Fallot:** RV-PA conduits may produce misleadingly low-pitched murmurs despite significant PR.

### 8.1 Diagnostic Pearls

- **PS Origin:** Congenital; rheumatic is rare.
- **Graham Steell:** Hallmark of PR.
- **RV Adaptation:** RV dysfunction occurs earlier in PS than LV in AS.

### 8.2 Therapeutic Pearls

- **Balloon Valvuloplasty:** For symptomatic moderate/severe PS or asymptomatic >64 mmHg.
- **Diuretics:** Use for right heart failure; avoid if PR is moderate to severe.
- **Transcatheter Replacement:** Option for severe PR after childhood repair.

---

REFERENCE TABLES

TABLE 278-1

**Causes of Pulmonic Valve Disease**

278 Pulmonic Valve Disease
Patrick T. O'Gara, Joseph Loscalzo

TABLE 278-1

**Causes of Pulmonic Valve Disease**

| VALVE LESION | ETIOLOGIES |
| --- | --- |
| **Pulmonic stenosis** | Congenital<br>Carcinoid<br>Tumor<br>Endocarditis |