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Pulmonary Stenosis: Diagnostic Imaging and Respiratory Care Perspectives

Reem Shelewh Alanzi [(1)] , Miad Azooz Musa Alsalhani [(1)] , Wesam Fayez Hammad Alshammari [(2)] , Amjad Sulaiman Saket Alshammary [(1)] , Fatimah Younis Ali Almohammedsaleh [(3)] , Shoroq Ameer Almohammed Saleh [(4)] , Mahdi Ali Alhelal [(5)] , Yasseen Jafar Alabdrabalnabi [(6)] , Haider Mohammed Almatar [(7)] , Ahmad Albaqshi [(4)]

(1) Buraidah Central Hospital, Buraidah, Ministry of Health, Saudi Arabia,
(2) King Fahd Specialist Hospital, Buraidah, Ministry of Health, Saudi Arabia,
(3) Al-Jabr Eye & ENT Hospital, Al-Ahsa, Ministry of Health, Saudi Arabia,
(4) Maternity and Children Hospital, Hail, Ministry of Health, Saudi Arabia,
(5) Hail General Hospital, Hail City, Ministry of Health, Saudi Arabia,
(6) King Salman Hospital, Hail, Ministry of Health, Saudi Arabia,
(7) Women and Maternity Hospital, Hail, Ministry of Health, Saudi Arabia

Abstract

Background: Pulmonary stenosis (PS) is a common congenital heart defect, accounting for 7-12% of cases, characterized by obstruction of the right ventricular outflow tract. It can occur in isolation or as part of complex syndromes. The clinical spectrum ranges from asymptomatic cases to severe presentations with heart failure.

Aim: This article provides a comprehensive review of PS, encompassing its etiology, pathophysiology, diagnostic evaluation, and contemporary management strategies to guide clinical practice.

Methods: A detailed narrative review was conducted, synthesizing current medical literature on PS. It examines diagnostic modalities, including echocardiography (the cornerstone), cardiac MRI, and CT, and analyzes evidence-based treatment guidelines from major cardiology societies.

Results: The severity of PS dictates management. Mild, asymptomatic cases often require only surveillance. For significant valvular PS, balloon pulmonary valvuloplasty is the preferred first-line intervention, yielding excellent outcomes, particularly for domed valves. Dysplastic valves or complex/subvalvular obstructions often necessitate surgical repair. Critical PS in neonates is a ductal-dependent emergency requiring prostaglandin therapy and urgent intervention. Long-term prognosis is generally favorable with appropriate treatment, though complications like arrhythmias, pulmonary regurgitation, and endocarditis can occur.

Conclusion: PS management requires accurate anatomical and hemodynamic assessment. Treatment is highly effective, ranging from monitoring to catheter-based or surgical interventions. A multidisciplinary, patient-tailored approach ensures optimal outcomes across the lifespan.

Keywords: Pulmonary stenosis, congenital heart disease, balloon valvuloplasty, echocardiography, right ventricular outflow tract obstruction.

Introduction

Pulmonary stenosis represents a relatively frequent congenital cardiac malformation and may present either as an isolated lesion or, more frequently, as a component of a more complex constellation of structural heart abnormalities, such as tetralogy of Fallot. The site of obstruction can vary along the right ventricular outflow tract, including subvalvular regions within the right ventricular infundibulum, the level of the pulmonary valve annulus and leaflets, or the supravalvular segments involving the main pulmonary artery and its proximal branches. This anatomic heterogeneity contributes to a broad clinical spectrum and influences both hemodynamic impact and therapeutic decision-making. Isolated pulmonary stenosis accounts for approximately 7% to 12% of congenital heart disease cases; however, in clinical practice it is more frequently encountered in association with other congenital anomalies, in which case it is present in nearly 25% to 30% of affected individuals [1]. The clinical manifestations of pulmonary stenosis are largely determined by the degree of obstruction to right ventricular outflow and the ability of the right ventricle to adapt to the resultant pressure overload. Patients with mild stenosis are often asymptomatic and may remain undiagnosed until adulthood, whereas those with moderate to severe obstruction are more likely to exhibit exertional dyspnea, reduced exercise tolerance, and fatigue, reflecting impaired forward flow and the limitations of

*Corresponding author e-mail: reemalanzi28@gmail.com (Reem Shelewh Alanzi).
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compensatory right ventricular hypertrophy. In a minority of cases, more alarming presentations such as exertional angina, presyncope, or even sudden cardiac arrest can occur, particularly when the hemodynamic burden is substantial or when coexistent coronary or myocardial abnormalities are present. Exceptionally, markedly dilated pulmonary artery aneurysms, which can arise in the context of chronically elevated post-stenotic pressures, may compress the left main coronary artery, thereby provoking myocardial ischemia and angina through an extrinsic mass effect rather than intrinsic coronary artery disease [1].

Accurate diagnosis and comprehensive anatomical and functional assessment are central to the management of pulmonary stenosis. Transthoracic echocardiography is the primary imaging modality and remains the cornerstone for initial evaluation, enabling visualization of valve morphology, estimation of pressure gradients using Doppler techniques, and assessment of right ventricular size and function. Nonetheless, in more complex cases, or when interventional or surgical procedures are contemplated, advanced cross-sectional imaging plays a pivotal adjunctive role. Cardiac computed tomography (CCT) offers high-resolution delineation of the pulmonary valve apparatus, right ventricular outflow tract, and pulmonary arterial tree, while cardiac magnetic resonance (CMR) provides superior quantification of right ventricular volumes, function, and flow dynamics, especially in patients with repaired congenital heart disease or prior interventions [2][3]. These modalities contribute crucial anatomic and hemodynamic details that refine risk stratification and procedural planning in candidates for intervention. Therapeutic strategies for pulmonary stenosis have evolved significantly with the expansion of catheter-based technologies. In contemporary practice, transcatheter approaches, particularly balloon pulmonary valvuloplasty, constitute the preferred first-line treatment for many patients with valvular pulmonary stenosis, owing to their favorable safety profile, efficacy in reducing transvalvular gradients, and shorter recovery times. Nevertheless, not all anatomic substrates are amenable to percutaneous intervention. Dysplastic valves, complex subvalvular or supravalvular obstructions, and certain forms of multilevel right ventricular outflow tract involvement may necessitate surgical repair or reconstruction. Thus, the ultimate choice between transcatheter and surgical therapy is influenced by the pattern and severity of obstruction, valve morphology, associated cardiac anomalies, and the overall clinical status of the patient. The degree of flow restriction across the pulmonary valve, together with detailed valve and outflow tract anatomy, guides therapeutic decision-making, in accordance with evidence-based recommendations from professional societies such as the American Heart Association (AHA) and the American College of Cardiology (ACC), which provide structured criteria for timing and modality of intervention in both pediatric and adult populations with congenital heart disease [3].

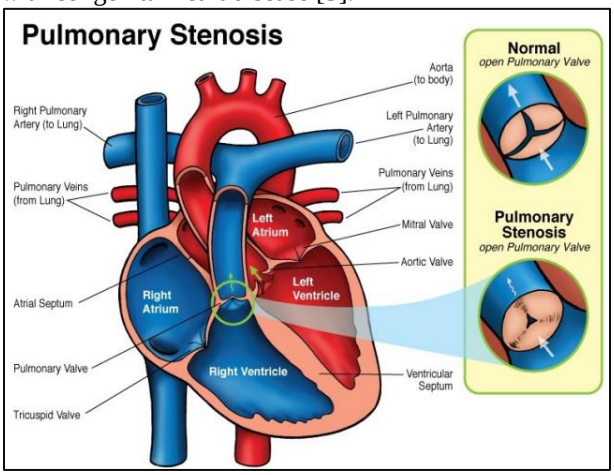

Fig. 1: Pulmonary Stenosis Vs. Normal Valve.

Etiology

Pulmonary stenosis arises from a heterogeneous group of etiologic mechanisms encompassing both congenital and acquired processes, and it may present either as an isolated valvular abnormality or in conjunction with more complex structural cardiac malformations. As an isolated lesion, valvular pulmonary stenosis results from intrinsic abnormalities of the pulmonary valve leaflets or annulus, which may be dysplastic, thickened, or fused, thereby limiting valve opening and impeding right ventricular outflow. However, pulmonary stenosis is frequently encountered in the context of congenital heart disease, where it forms an integral component of more complex anatomical configurations. It is classically associated with tetralogy of Fallot, in which right ventricular outflow tract obstruction constitutes one of the defining features, but it may also accompany tricuspid atresia, complete transposition of the great arteries, congenitally corrected transposition, and double outlet right ventricle. In these settings, the presence of pulmonary stenosis may significantly influence intracardiac hemodynamics, ventricular loading conditions, and the selection and timing of surgical or interventional strategies. Genetic factors play a central role in the pathogenesis of many forms of pulmonary stenosis, particularly when it occurs in association with defined syndromic conditions. Noonan syndrome is among the best-characterized genetic syndromes linked to valvular pulmonary stenosis. This multisystem disorder is most commonly associated with pathogenic variants in the PTPN11 gene, but mutations in other components of the RAS–MAPK signaling pathway, including KRAS, SOS1, and RAF1, have also been identified as causative and contribute to the phenotypic spectrum [4][5][6][7]. In individuals with Noonan syndrome, pulmonary valve dysplasia and stenosis are highly prevalent and may coexist with other cardiac anomalies such as

hypertrophic cardiomyopathy. Peripheral pulmonary stenosis, in which the narrowing predominantly affects the branch pulmonary arteries rather than the valve itself, constitutes another important phenotypic expression of genetically mediated arteriopathy. This pattern is frequently observed in Alagille syndrome, a disorder characterized by cholestatic liver disease, characteristic facial features, and vertebral anomalies, which is most often caused by mutations in the JAG1 gene on chromosome 12q24, and less commonly by alterations in NOTCH2 [8]. Similarly, Williams–Beuren syndrome, a neurodevelopmental and connective tissue disorder associated with an elastin (ELN) gene mutation on chromosome 7q11.23, is classically linked to supravalvular aortic stenosis but may also present with peripheral pulmonary artery stenosis, thereby highlighting the shared molecular underpinnings of vascular stenoses in elastin-related arteriopathies [9].

Not all etiologies of pulmonary stenosis are inherited; some result from environmental or infectious exposures during critical periods of fetal development. Maternal rubella infection during pregnancy is a well-recognized cause of congenital heart disease within the broader constellation of congenital rubella syndrome. In this context, valvular pulmonary stenosis may arise as part of a spectrum of cardiac malformations that also includes patent ductus arteriosus and septal defects. Although the lesion itself is congenital, the underlying mechanism is teratogenic rather than genetic, reflecting direct viral damage and disruption of normal organogenesis rather than heritable mutations [10][11]. The declining incidence of maternal rubella syndrome in regions with robust vaccination programs underscores the importance of public health measures in modulating the prevalence of certain forms of pulmonary stenosis. In some instances, previously unrecognized pulmonary stenosis may first become clinically apparent during states of increased hemodynamic demand, such as pregnancy. The physiologic expansion of plasma volume and cardiac output associated with gestation can unmask latent right ventricular outflow obstruction in women with undiagnosed valvular lesions, leading to the emergence or worsening of symptoms such as exertional dyspnea, fatigue, and, less commonly, syncope or arrhythmias [12]. Additionally, valvular involvement by carcinoid heart disease can give rise to acquired pulmonary stenosis, particularly in patients with carcinoid tumors that secrete vasoactive substances capable of inducing fibrotic thickening and retraction of right-sided heart valves. In this setting, pulmonary stenosis may develop de novo or exacerbate preexisting obstruction, and it may coexist with tricuspid regurgitation as part of a characteristic right-sided valvulopathy [13]. Beyond congenital and carcinoid-related mechanisms, acquired pulmonary stenosis may also develop as a complication of other cardiac or systemic conditions. Rheumatic heart disease, although far more commonly associated with mitral and aortic valve involvement, can occasionally affect the pulmonary valve, leading to commissural fusion, leaflet thickening, and progressive stenosis. Similarly, prior cardiothoracic surgeries involving the right ventricular outflow tract or pulmonary valve—such as repairs for tetralogy of Fallot, pulmonary valve replacement, or conduit placement—may predispose to restenosis due to scarring, calcification, or degeneration of prosthetic material. Cardiac and mediastinal tumors, including pericardial sarcoma, teratoma, thymoma, and malignancies associated with Hodgkin disease, may also produce functional pulmonary stenosis through direct invasion, external compression of the right ventricular outflow tract, or distortion of the pulmonary valve apparatus [14]. Collectively, these diverse etiologic pathways underscore the need for a detailed clinical, genetic, and imaging evaluation in patients with pulmonary stenosis, both to elucidate the underlying cause and to guide individualized management and surveillance strategies [13][14].

Epidemiology

Pulmonary stenosis represents a notable proportion of congenital cardiac malformations, with isolated valvar pulmonary stenosis accounting for approximately 7% to 12% of all congenital heart disease cases, making it one of the more frequently observed obstructive lesions within pediatric and adult congenital cardiology [15]. Although many patients present with isolated valvar involvement, a substantial proportion exhibit broader systemic manifestations or associated developmental abnormalities. Extracardiac and neurodevelopmental comorbidities are reported in nearly 56% of individuals diagnosed with pulmonary stenosis, a statistic that underscores the importance of comprehensive clinical evaluation and the strong possibility of underlying genetic contributions to the phenotype. When these comorbidities are present, the likelihood of obtaining a definitive molecular diagnosis is significantly higher, reflecting the intersection between cardiac malformations and genetic syndromic conditions. One of the most well-recognized genetic associations is the link between valvar pulmonary stenosis and Noonan syndrome, a multisystem disorder frequently caused by pathogenic variants affecting the RAS–MAPK signaling pathway. Among these, *PTPN11* mutations represent the predominant molecular abnormality, identified in approximately 50% of patients with Noonan syndrome who also have pulmonary stenosis [7][16]. The high frequency of pulmonary valve dysplasia in this population highlights the importance of targeted genetic testing when characteristic facial features, developmental delays, or other systemic findings point toward a syndromic etiology. These genotype–phenotype correlations have not only enhanced diagnostic accuracy but also provided insights into the developmental pathways governing valvar morphogenesis [7][16].

Beyond syndromic associations, pulmonary stenosis also occurs in familial clusters that lack overt extrinsic features, suggesting the presence of inherited nonsyndromic forms. Emerging evidence indicates that mutations in transcription factors crucial for cardiac embryogenesis, particularly *GATA4*, may contribute to these familial presentations. *GATA4* plays a central role in myocardial and valvar development, and its disruption may lead to a spectrum of congenital cardiac defects, including pulmonary stenosis. Although this hereditary pattern appears less common than syndromic forms, its recognition has implications for genetic counseling and screening of first-degree relatives. In contrast to several congenital cardiac disorders that present with distinct gender disparities, pulmonary stenosis does not demonstrate a meaningful predilection for either sex, occurring with comparable frequency in males and females [17]. This balanced distribution further suggests that environmental or epigenetic influences may be less significant contributors to disease onset compared with intrinsic genetic and developmental mechanisms. Overall, the epidemiological landscape of pulmonary stenosis reflects a complex interplay between isolated anatomical defects, syndromic conditions, and familial genetic predispositions, underscoring the necessity of integrated clinical, genetic, and imaging approaches to optimize diagnosis, counseling, and long-term management [17].

Pathophysiology

Pulmonary stenosis encompasses a spectrum of obstructive lesions affecting the right ventricular outflow tract and is traditionally classified according to the anatomic level at which the obstruction occurs. Three principal types are recognized—valvular, subvalvular, and supravalvular—each with distinct structural characteristics but similar hemodynamic consequences: an increased resistance to right ventricular ejection, elevation of right ventricular systolic pressure, and the development of compensatory right ventricular hypertrophy. The functional severity of the obstruction may range from mild to critical, and this gradation plays a central role in determining clinical presentation, timing of intervention, and long-term prognosis [18][3]. Valvular pulmonary stenosis is the most frequent form and typically reflects congenital maldevelopment of the pulmonary valve apparatus. In its classic form, the pulmonary valve leaflets are thin but exhibit varying degrees of fibrosis, thickening, and commissural fusion. These morphological alterations prevent normal systolic opening and create a narrowed, resistant orifice through which blood must be ejected. As a consequence, the valve assumes a characteristic domed or conical configuration during systole, with a small central opening serving as the effective outflow channel [19]. This valvular subtype is most often identified in childhood, usually as part of routine evaluation for a cardiac murmur or exertional symptoms. Although pulmonary valve stenosis is predominantly congenital, rare acquired cases may arise in adulthood due to conditions such as rheumatic heart disease, which can induce leaflet thickening and commissural fusion, or carcinoid syndrome, in which serotonin-mediated fibrotic deposition distorts the valve structure [19]. Within valvular pulmonary stenosis, there is considerable morphological variability that has important implications for both pathophysiology and therapeutic options. In many patients, the valve leaflets are thin but partially fused at their commissures, leading to a doming pattern without gross leaflet dysplasia. In contrast, a dysplastic pulmonary valve is characterized by markedly thickened, stiff cusps with little or no commissural fusion, resulting in a rigid and often hypoplastic annulus. This dysplastic variant is notably prevalent in patients with Noonan syndrome, who frequently display a small pulmonary annulus and hypoplastic proximal pulmonary arteries, thereby compounding the obstruction to right ventricular outflow [20]. Additionally, the number and configuration of pulmonary valve leaflets may vary. Bicuspid pulmonary valves, although less common than bicuspid aortic valves, are more frequently encountered in association with tetralogy of Fallot. Even more atypical patterns, such as quadricuspid valves, have been reported and can alter flow dynamics across the right ventricular outflow tract [20].

Subvalvular, or infundibular, pulmonary stenosis describes obstruction that arises below the pulmonary valve, within the muscular outflow tract of the right ventricle. In this setting, hypertrophy and abnormal alignment of the infundibular muscle bundles create a narrowed channel proximal to the valve. This variant is classically seen in conditions such as tetralogy of Fallot, in which anterior deviation of the infundibular septum contributes to right ventricular outflow tract obstruction, and in the double-chambered right ventricle, where anomalous muscle bundles partition the ventricular cavity into high- and low-pressure chambers. Subvalvular stenosis is also observed in 20% to 30% of patients with Noonan syndrome who have hypertrophic cardiomyopathy; in these individuals, marked right ventricular hypertrophy accentuates narrowing of the outflow tract [7]. Importantly, subvalvular obstruction may be primary or secondary. In some patients with longstanding valvular pulmonary stenosis, chronic pressure overload leads to compensatory right ventricular hypertrophy, which can secondarily encroach on the outflow tract and generate a dynamic infundibular gradient. When the primary valvular obstruction is relieved through valvotomy or balloon valvuloplasty, the stimulus for hypertrophy diminishes, and the secondary subvalvular component often regresses over time, resulting in improved

hemodynamics and reduced right ventricular pressure burden [21][22]. Supravalvular pulmonary stenosis, frequently referred to as peripheral pulmonary stenosis, involves obstruction located above the pulmonary valve, either within the main pulmonary artery, its branch vessels, or both. The narrowing may be focal, multifocal, or diffuse, and it significantly alters pulmonary blood flow distribution. This form is frequently associated with complex congenital heart defects such as tetralogy of Fallot, double-outlet right ventricle, and transposition of the great arteries, in which abnormalities of conotruncal development affect both the great vessels and their branches [23][24][25][26][27][28]. Supravalvular pulmonary stenosis is also a well-recognized feature of several genetic syndromes, including Noonan, Williams, and Alagille syndromes, wherein intrinsic arterial wall pathology, elastin abnormalities, or Notch pathway defects result in diffuse arteriopathy and segmental stenoses of the pulmonary arterial tree [23][24][25][26][27][28]. In addition, supravalvular stenosis may develop as a postoperative complication, for example following surgical repair of transposition of the great arteries, where surgical manipulation, scarring, or anastomotic narrowing at the level of the pulmonary arteries can lead to iatrogenic obstruction [29].

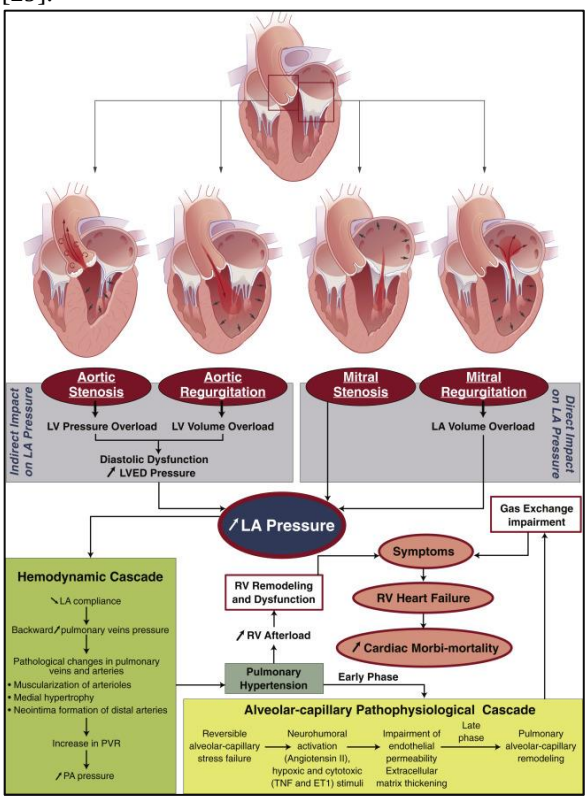

Fig. 2: Pathophysiology of pulmonary stenosis.

The hemodynamic severity of pulmonary stenosis, irrespective of its anatomic level, is customarily quantified using Doppler echocardiography or invasive catheterization. These modalities assess the peak pressure gradient across the obstructed segment, which serves as a surrogate for the degree of flow limitation. Mild stenosis is usually defined by a peak gradient of less than 36 mm Hg, a range in which right ventricular pressure is only modestly elevated and symptoms are typically absent or minimal. When the peak gradient rises into the range of 36 to 64 mm Hg, the lesion is classified as moderate, often corresponding to a greater degree of right ventricular hypertrophy and a higher likelihood of exertional symptoms. Severe stenosis is characterized by a peak gradient exceeding 64 mm Hg, reflecting a substantial impediment to right ventricular outflow and a marked increase in right ventricular systolic pressure relative to the pulmonary artery [3]. Over time, this degree of obstruction can lead to progressive right ventricular dysfunction, tricuspid regurgitation, arrhythmias, and diminished exercise capacity. At the extreme end of the spectrum lies critical pulmonary stenosis, which is most often encountered in neonates and young infants. In this scenario, the pulmonary valve or annulus is so severely narrowed that antegrade blood flow from the right ventricle into the pulmonary artery is profoundly compromised. As a result, adequate pulmonary perfusion becomes heavily dependent on right-to-left or left-to-right shunting through fetal channels such as the ductus arteriosus. In these infants, maintenance of ductal patency is lifesaving; prostaglandin-E1 infusion is administered to prevent ductal closure and preserve pulmonary blood flow until definitive intervention—typically balloon valvuloplasty or surgical valvotomy—can be undertaken [18]. The pathophysiology of critical stenosis therefore involves not only the obstructed valve itself but also the transitional circulation of the newborn, in which rapid changes in pulmonary vascular resistance and ductal dynamics can precipitate profound hypoxemia and hemodynamic instability if not promptly addressed. Across all types and severities, the central pathophysiologic theme of pulmonary stenosis is the creation of a pressure gradient between the right ventricle and the pulmonary artery, which in turn drives right ventricular hypertrophy and alters ventricular–vascular coupling. The precise anatomic site of the narrowing, its extent, and associated syndromic or structural heart disease modify the clinical expression, long-term course, and response to intervention. Understanding these nuances is essential for tailoring imaging assessments, selecting the optimal therapeutic approach, and anticipating potential complications over the lifespan of patients with this diverse and complex congenital lesion [18].

History and Physical

Clinical Features

The clinical presentation of pulmonary stenosis varies widely and is fundamentally shaped by both the anatomical severity of the obstruction and the right ventricle's ability to compensate for the increased outflow resistance. Many individuals with mild pulmonary stenosis remain entirely asymptomatic throughout childhood and adulthood, as

the hemodynamic burden imposed by a minimal right ventricular outflow gradient is often well tolerated. These patients are frequently diagnosed incidentally during routine clinical examinations when a characteristic murmur prompts further evaluation. In contrast, those with moderate or severe pulmonary stenosis are more likely to manifest symptoms resulting from elevated right ventricular systolic pressures and impaired forward flow. Dyspnea on exertion and exertional fatigue represent the most common symptomatic complaints, reflecting the diminished capacity of the hypertrophied right ventricle to augment cardiac output during physical activity. The degree of symptomatology often correlates with the magnitude of the pressure gradient across the pulmonary valve and the degree to which the right ventricle has undergone adaptive hypertrophic remodeling [1]. Although less frequent, more dramatic manifestations may occur. Chest pain resembling angina has been reported in some patients, typically associated with severe obstruction. This symptom may be related to subendocardial ischemia from markedly elevated right ventricular pressures; however, it may also be produced by external compression of the left main coronary artery when a substantial post-stenotic pulmonary artery aneurysm is present. In exceedingly rare scenarios, individuals may present with sudden cardiac arrest, particularly in the context of extreme obstruction or ventricular arrhythmias precipitated by advanced right ventricular hypertrophy or ischemia [30]. Physical examination plays a pivotal role in the initial assessment of pulmonary stenosis, and characteristic auscultatory findings provide valuable clues regarding severity. In mild pulmonary stenosis, the cardiac examination typically reveals a normal first heart sound (S1) followed by a high-frequency ejection click, which is produced by the abrupt doming of the mobile but stenotic pulmonary valve leaflets during early systole. The intensity of the pulmonic component of the second heart sound (P2) may range from normal to slightly accentuated, reflecting minimal compromise of pulmonary blood flow. As the obstruction becomes more pronounced, the auscultatory profile evolves. In moderate pulmonary stenosis, S1 remains audible, but the ejection click tends to approximate S1 more closely due to rising right ventricular pressures. A systolic murmur of ejection quality becomes more prominent, and the second heart sound (S2) demonstrates splitting, with a softened P2 component. The degree of splitting is influenced by the severity of obstruction and delayed pulmonic valve closure [31].

In severe pulmonary stenosis, the ejection click may disappear entirely, as the markedly stiff pulmonary valve leaflets may fail to produce an audible sound. The systolic ejection murmur increases in intensity, often becoming harsh and widely radiating. The second heart sound is widely split due to significantly delayed closure of the pulmonic valve, and P2 may be extremely soft or completely inaudible as pulmonary blood flow diminishes [31]. In extreme cases, the murmur may radiate to the back, reflecting turbulent flow through a narrowed right ventricular outflow tract [32]. Additionally, reduced right ventricular compliance in advanced disease can result in the presence of an audible fourth heart sound (S4), indicative of forceful atrial contraction against a stiff, hypertrophied right ventricle [33]. Infants born with critical pulmonary stenosis constitute a distinct and potentially life-threatening presentation. These neonates often develop cyanosis shortly after birth due to inadequate antegrade pulmonary blood flow. The resultant elevation in right atrial pressure promotes right-to-left shunting through a patent foramen ovale or an atrial septal defect, contributing to systemic desaturation. Signs of systemic venous congestion may also emerge, including hepatomegaly, peripheral edema, and poor perfusion, driven by right ventricular dysfunction and impaired venous return [18]. Progressive right ventricular dilation in infants or older individuals may signal the presence of an atrial septal defect, while sustained increases in right atrial pressure can exacerbate intracardiac shunting and further compromise oxygenation. In severe or long-standing cases of pulmonary stenosis, systemic venous congestion may become prominent, manifesting as jugular venous distention, hepatomegaly, and peripheral edema [30]. In children and adults with moderate or severe obstruction, additional physical findings can offer diagnostic insight. A left parasternal heave is often appreciated, reflecting right ventricular hypertrophy secondary to chronic pressure overload. The characteristic systolic ejection murmur frequently radiates posteriorly, and its quality may evolve as the outflow gradient worsens [32]. Auscultation of an S4, while not universal, is indicative of markedly reduced right ventricular compliance and is more often associated with severe, long-standing obstruction [33]. Altogether, the history and physical examination provide indispensable early diagnostic information, guide the need for further imaging, and assist in determining the urgency of intervention [32][33].

Evaluation

The evaluation of pulmonary stenosis relies primarily on imaging and hemodynamic assessment to determine the anatomic location of obstruction, quantify its severity, and identify associated structural or functional abnormalities. Echocardiography remains the cornerstone of diagnostic evaluation, offering a noninvasive, widely available, and reliable modality capable of delineating valvular morphology, right ventricular outflow tract anatomy, and hemodynamic gradients with high fidelity. In most clinical settings, a transthoracic echocardiogram provides sufficiently detailed visualization of the pulmonary valve, allowing for assessment of leaflet mobility, commissural fusion, and the presence of post-stenotic dilation of the pulmonary artery.

Transthoracic echocardiography also enables real-time evaluation of right ventricular size, wall thickness, systolic function, and the presence of coexisting congenital anomalies, thereby furnishing a comprehensive overview of cardiac structure and performance [34][35]. Despite its utility, transthoracic echocardiography may occasionally offer suboptimal imaging windows, particularly in adults or patients with complex cardiac anatomy. In such cases, transesophageal echocardiography (TEE) may be employed to obtain higher-resolution images. TEE is particularly valuable when endocarditis is suspected, as it offers superior visualization of vegetations, valve thickening, and complications such as abscess formation, which may influence management decisions [36][37]. Although echocardiography forms the foundation of diagnostic evaluation, electrocardiography provides complementary information, particularly regarding the electrical manifestations of right ventricular hypertrophy. The degree of electrocardiographic abnormality generally correlates with the severity of obstruction. Mild pulmonary stenosis may manifest only subtle changes, such as right axis deviation, whereas more advanced cases often reveal pronounced R waves in the right precordial leads, particularly V1 and aVR, indicative of significant right ventricular hypertrophy. P-wave enlargement may also appear, reflecting increased right atrial pressure and atrial dilation secondary to impaired right ventricular compliance [38].

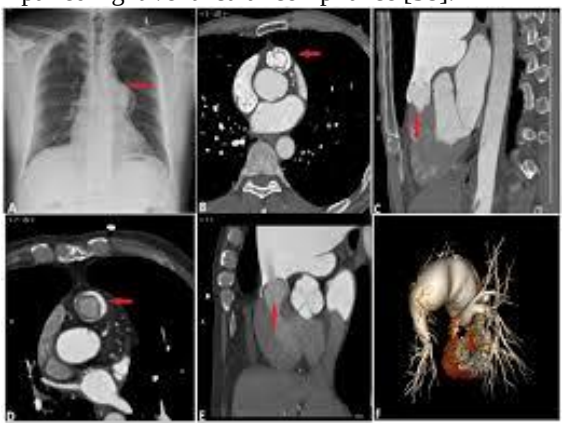

Fig. 3: CT Scan of Pulmonary Stenosis.

Doppler echocardiographic studies serve an essential role in quantifying the severity of pulmonary stenosis by measuring flow velocities and generating pressure gradients across the pulmonary valve. These gradients provide a physiologic estimate of the functional burden imposed by the obstruction and are integral to therapeutic decision-making. Major cardiovascular societies, including the European Association of Echocardiography, the American Society of Echocardiography, the American Heart Association (AHA), the American College of Cardiology (ACC), and the European Society of Cardiology (ESC), classify stenosis severity using standardized Doppler thresholds. Mild stenosis corresponds to a peak Doppler gradient of less than 36 mm Hg or a jet velocity under 3 m/s. Moderate stenosis is defined by a gradient between 36 and 64 mm Hg or a velocity of 3 to 4 m/s. Severe stenosis is characterized by a peak gradient exceeding 64 mm Hg or a velocity greater than 4 m/s [34][35][39]. These hemodynamic classifications guide clinical management, particularly when determining the need for intervention or monitoring progression over time. Although echocardiography is usually sufficient for both diagnosis and severity assessment, additional imaging modalities may be required in select clinical circumstances. Cardiac catheterization, once the gold standard for evaluating pulmonary stenosis, is now seldom necessary for diagnostic purposes due to advancements in noninvasive imaging. Nonetheless, catheter-based assessment remains valuable when percutaneous valvuloplasty is planned, as it provides the opportunity for direct hemodynamic measurement and interventional treatment during the same procedure. Pulmonary angiography, similarly, is rarely used solely for diagnostic evaluation but may assist in delineating branch pulmonary artery anatomy in complex congenital conditions [34][35].

Cardiac magnetic resonance (CMR) has emerged as a highly useful adjunctive modality, particularly in patients with poor echocardiographic windows, repaired congenital heart disease, or suspected complex right ventricular outflow tract pathology. CMR yields precise measurements of right ventricular volumes, ejection fraction, and myocardial mass while offering superior assessment of pulmonary artery size, anatomy, and flow distribution. It is also valuable in distinguishing primary valvular disease from subvalvular or supravalvular forms of stenosis and can quantify regurgitant fractions in patients with prior valvotomy or valvuloplasty [40][41]. Furthermore, fetal CMR can complement fetal echocardiography in prenatal evaluations by providing enhanced visualization of pulmonary artery branches and right ventricular outflow tract configurations in utero, aiding in delivery planning and parental counseling. Cardiac computed tomography (CCT) serves as an alternative imaging tool when CMR is contraindicated, such as in patients with metallic implants or severe claustrophobia. CCT offers excellent spatial resolution and is especially useful for evaluating the pulmonary arteries, detecting supravalvular stenosis, and visualizing calcification or structural abnormalities of the pulmonary valve. However, because CCT involves ionizing radiation and iodinated contrast, its use is typically reserved for cases in which CMR is not feasible [3]. By contrast, plain chest radiography lacks the sensitivity needed to diagnose pulmonary stenosis definitively. Nevertheless, it may reveal supportive findings, such as prominence of the main pulmonary artery segment due to post-stenotic dilation or an enlarged right heart border reflecting right ventricular hypertrophy. Although nonspecific, these findings may prompt further diagnostic evaluation with echocardiography

in the appropriate clinical context [30]. Collectively, the evaluation of pulmonary stenosis relies on a structured integration of clinical examination, electrocardiographic findings, and multimodal imaging, with echocardiography positioned at the center of diagnostic decision-making. This comprehensive approach ensures accurate characterization of the lesion, facilitates timely intervention, and informs long-term management strategies tailored to the individual patient [3][30].

Treatment / Management

The management of pulmonary stenosis is fundamentally guided by the severity of obstruction across the right ventricular outflow tract, particularly at the level of the pulmonary valve, and by the anatomical characteristics of the valve and adjacent structures. Contemporary treatment strategies are anchored in evidence-based recommendations issued by major professional societies, notably the American Heart Association (AHA) and the American College of Cardiology (ACC), which have periodically revised and updated their guidelines to reflect advances in diagnostic imaging, catheter-based interventions, and surgical techniques [36]42. A rational approach to care begins with careful stratification of patients according to the hemodynamic burden of the stenosis, symptom status, and the presence of associated cardiac or extracardiac conditions. For individuals with mild obstruction, particularly those who are asymptomatic, conservative management with structured surveillance is generally appropriate. The AHA and ACC propose specific follow-up intervals based on Doppler-derived gradients. Asymptomatic patients whose peak Doppler gradient across the pulmonary valve is less than 30 mm Hg can be monitored relatively infrequently, with clinical review, electrocardiography, and Doppler echocardiography approximately every five years. This long interval reflects the typically benign natural history of very mild stenosis and the low likelihood of rapid progression in this subgroup. In contrast, asymptomatic patients whose peak Doppler gradient exceeds 30 mm Hg warrant closer observation, with follow-up Doppler echocardiography recommended every two to five years, since these patients are at greater risk of developing symptoms or progressive right ventricular hypertrophy over time [36][42]. These surveillance strategies aim to identify the transition from compensated to clinically significant obstruction before the onset of irreversible right ventricular dysfunction.

When intervention becomes necessary, balloon pulmonary valvuloplasty is the preferred first-line treatment for most patients with valvular pulmonary stenosis, particularly when the valve is of the classic domed, non-dysplastic type. This catheter-based procedure involves the advancement and inflation of a balloon across the stenotic valve to disrupt commissural fusion and enlarge the effective valve orifice, thereby reducing the transvalvular gradient. The AHA and ACC recommend balloon valvuloplasty in several specific clinical scenarios. Asymptomatic patients with a domed pulmonary valve and a peak Doppler gradient greater than 60 mm Hg are considered appropriate candidates, even in the absence of symptoms, because such high gradients are associated with a substantial long-term risk of right ventricular strain and adverse outcomes if left untreated. In symptomatic patients who present with unexplained heart failure, cyanosis due to interatrial right-to-left shunting, or exercise intolerance, balloon valvuloplasty is advised when they have a domed pulmonary valve with a peak Doppler gradient exceeding 50 mm Hg or a mean Doppler gradient greater than 30 mm Hg, as these hemodynamic thresholds typically correlate with clinically impactful obstruction [36][42]. The situation is more nuanced in patients with dysplastic pulmonary valves. Dysplastic valves are often thickened and rigid, with minimal commissural fusion, and they respond less favorably to balloon dilation than domed valves. Consequently, surgical intervention is usually preferred for this anatomic subtype. Nonetheless, balloon valvuloplasty may still be considered reasonable in selected patients with dysplastic valves. In asymptomatic individuals with a dysplastic pulmonary valve, intervention can be contemplated when the peak Doppler gradient exceeds 60 mm Hg or the mean Doppler gradient is above 40 mm Hg. In symptomatic patients with dysplastic valves, the procedure may be offered when the peak gradient is greater than 50 mm Hg or the mean gradient exceeds 30 mm Hg [36]. These criteria reflect the balance between the potential benefits of gradient reduction and the limitations imposed by the abnormal valve morphology [42].

It is important to recognize that even when technically successful, balloon valvuloplasty does not always provide a permanent solution. Restenosis can occur, and some patients may require reintervention during long-term follow-up. In addition, the procedure can predispose to varying degrees of pulmonary regurgitation due to disruption of valve coaptation. Risk factors associated with a higher likelihood of significant post-procedural regurgitation include very young age, low body weight at the time of intervention, a small pulmonary annulus diameter, high baseline systolic gradient, an elevated right ventricular-to-systemic pressure ratio, and the presence of very severe pulmonary stenosis pre-intervention [43]. Understanding these predictors helps in counseling patients and families regarding expectations and potential long-term sequelae. There are also clear situations in which balloon valvuloplasty is not recommended. Asymptomatic patients with normal cardiac output and a Doppler peak instantaneous gradient below 50 mm Hg generally do not benefit from the procedure, given the relatively low risk posed by such gradients and the potential complications of an invasive intervention. Similarly,

balloon valvuloplasty is not advised in symptomatic patients with pulmonary stenosis who already have severe pulmonary regurgitation, as further disruption of valve integrity would likely exacerbate regurgitation and worsen right ventricular volume overload. The procedure is also inappropriate in symptomatic patients whose Doppler peak instantaneous gradient is less than 30 mm Hg, since their symptoms are unlikely to be explained by mild stenosis alone and alternative diagnoses should be considered [36][42][43].

Surgical intervention retains a vital role in the management of pulmonary stenosis, particularly when catheter-based options are unsuitable or have failed. Surgical valvotomy or valve repair is indicated in patients with moderate-to-severe valvular stenosis who are symptomatic and either not candidates for balloon valvuloplasty or have persistent significant gradients following an attempted catheter-based procedure. Surgery is also the treatment of choice in patients with severe valvular stenosis accompanied by severe pulmonary regurgitation, where comprehensive valve reconstruction or replacement may be necessary to address both obstruction and regurgitation. Additional indications include the presence of a markedly hypoplastic pulmonary annulus, in which balloon dilation alone would be insufficient, and the presence of subvalvular or supravalvular stenosis, where obstruction lies beyond the reach of simple valvuloplasty. Dysplastic pulmonary valves associated with severe tricuspid regurgitation may also require surgical correction, particularly if a Maze procedure is planned for concomitant atrial arrhythmias. Furthermore, if a patient is already undergoing cardiac surgery for another indication—such as repair of a concomitant congenital defect—correction of significant pulmonary stenosis during the same procedure is often appropriate [36]. For patients with supravalvular or subvalvular pulmonary stenosis, particularly when the obstruction involves discrete or segmental narrowing of the pulmonary arteries or right ventricular outflow tract, percutaneous pulmonary artery balloon angioplasty with or without stent placement offers an acceptable and often effective therapeutic option. This approach can relieve stenosis at levels above or below the valve and improve pulmonary blood flow distribution, especially in complex congenital heart disease or after prior surgical repairs. The decision to use stents depends on lesion characteristics, vessel size, and the anticipated need for future growth or reintervention [36][42][43].

An additional dimension of management concerns the prevention of infective endocarditis, especially in individuals with residual lesions or prosthetic material. Antibiotic prophylaxis prior to dental procedures and vaginal delivery is considered reasonable in selected patients with pulmonary stenosis. These include those with a prosthetic cardiac valve or prosthetic material used for valve repair, individuals with a history of prior infective endocarditis, and patients who have other cyanotic congenital cardiac lesions, including those with systemic-to-pulmonary shunts or conduits. In general, antibiotic prophylaxis is recommended for patients with a documented history of infective endocarditis, for those with mechanical or biological prosthetic valves, for individuals who have received intracardiac prosthetic material within the preceding six months, for those with residual intracardiac shunts in or adjacent to prosthetic material, and for patients with cyanotic congenital heart disease. In contrast, prophylaxis is not recommended for routine non-dental procedures, such as esophagogastroduodenoscopy or colonoscopy, in the absence of active infection, as the risk of bacteremia in these contexts is low and does not justify antibiotic exposure [30]. Management becomes particularly urgent and complex in the setting of critical pulmonary stenosis in the newborn. In this extreme form, the lesion approximates pulmonary atresia, creating a ductal-dependent circulation in which survival is contingent upon maintaining patency of the ductus arteriosus to ensure adequate pulmonary blood flow. As the ductus arteriosus begins to close in the early postnatal period, these neonates develop progressive hypoxemia and worsening cyanosis, rapidly becoming hemodynamically unstable if left untreated. Prostaglandin E1 infusion is therefore initiated promptly to prevent ductal closure or to reopen a constricting ductus, thereby re-establishing a conduit for pulmonary perfusion. In some cases, ductal stenting may be employed as a more durable means of preserving ductal flow [18][44].

The intracardiac anatomy in critical pulmonary stenosis often includes a severely hypertrophied, restrictive, and hypoplastic right ventricle, particularly when no ventricular septal defect is present. Under these circumstances, the right ventricle may be incapable of generating sufficient forward flow to support the pulmonary circulation, and the cardiac output is effectively dependent on blood mixing at the atrial level. A patent foramen ovale or a small atrial septal defect allows systemic and pulmonary venous return to mix and pass into the subaortic ventricle, which functions as the primary systemic pumping chamber. To optimize this mixing and decompress the right atrium, enlargement of the interatrial communication is often necessary. This is accomplished via atrial septostomy, commonly performed as a Rashkind balloon procedure in the cardiac catheterization laboratory or, when required, at the bedside in the cardiac intensive care unit under ultrasound guidance with or without fluoroscopy. Static balloon dilation or atrial stent placement can be used to further ensure adequate and lasting interatrial communication [18][44]. Definitive anatomic repair may not be immediately feasible in neonates with severely hypoplastic right ventricles. In such cases, a strategy of staged single-ventricle palliation is frequently adopted, in which surgical procedures are

performed sequentially to redirect venous return and optimize systemic and pulmonary blood flow. However, in some infants whose right ventricle is small but capable of supporting a portion of the cardiac output, a one-and-a-half ventricle repair may be considered as the initial palliative operation. In this configuration, part of the systemic venous blood is directed to the pulmonary arteries via a cavopulmonary connection (such as a bidirectional Glenn shunt), while the right ventricle continues to pump some blood into the pulmonary circulation. This approach has advantages over a classic Fontan pathway, as it preserves pulsatile flow to the lungs and may reduce the risk of complications such as the development of pulmonary arteriovenous malformations, which can occur after Glenn or Fontan procedures [45][46]. Over time, if the right ventricle demonstrates adequate growth and functional capacity, conversion to a full biventricular repair may become possible, transforming a physiologically single-ventricle circulation into a two-ventricle system [45][46].

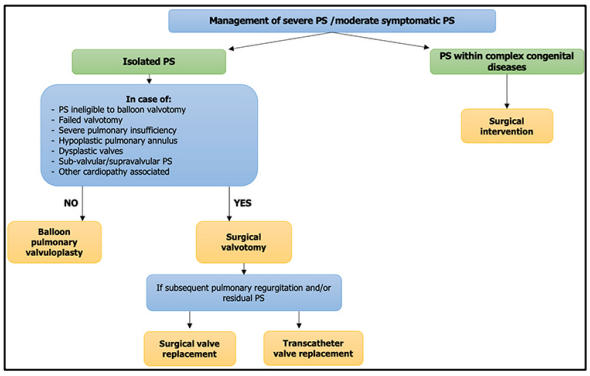

Fig. 4: Management of pulmonary stenosis.

Recent advances in fetal intervention have expanded the therapeutic possibilities for the most severe forms of right ventricular outflow tract obstruction. In selected fetuses with critical pulmonary stenosis or pulmonary valve atresia, in-utero pulmonary valvuloplasty has been performed with the goal of promoting right ventricular growth before birth [47]. By relieving the obstruction during fetal life, this procedure may allow the right ventricle to develop more normally and increase the likelihood that the infant will be a candidate for postnatal biventricular repair, rather than requiring lifelong single-ventricle palliation. Although technically demanding and limited to specialized centers, fetal intervention exemplifies the evolving, proactive philosophy in managing complex congenital heart disease. In summary, the treatment and management of pulmonary stenosis span a broad continuum, from conservative surveillance in asymptomatic patients with mild gradients to sophisticated catheter-based and surgical interventions in those with severe or critical disease. Decisions are tailored according to valve morphology, gradient severity, right ventricular function, associated anomalies, and patient age, all within the framework of guideline-directed care provided by organizations such as the AHA and ACC [36]. Through careful integration of noninvasive imaging, hemodynamic assessment, interventional techniques, and surgical expertise, clinicians can optimize outcomes for patients with this diverse and potentially complex congenital lesion across the lifespan.

Differential Diagnosis

The differential diagnosis of pulmonary stenosis spans a wide range of congenital and acquired conditions, reflecting the diverse mechanisms by which obstruction to pulmonary blood flow can occur. In infants, careful differentiation is essential because early recognition of specific structural cardiac abnormalities can significantly influence management strategies, surgical timing, and long-term outcomes. One of the key considerations is distinguishing isolated valvular pulmonary stenosis from congenital heart defects in which pulmonary stenosis appears as part of a broader anatomic complex. Conditions such as double-chambered right ventricle and double-outlet right ventricle can produce right ventricular outflow tract obstruction that mimics or coexists with pulmonary valvular disease. In a double-chambered right ventricle, anomalous muscular bundles divide the right ventricular cavity, creating a pressure gradient proximal to the pulmonary valve, whereas double-outlet right ventricle results in misalignment of the outflow tracts that can be accompanied by various degrees of subpulmonic obstruction. Absent pulmonary valve, a variant often associated with tetralogy of Fallot, is another important differential consideration. In this condition, massively dilated pulmonary arteries and a rudimentary valve apparatus produce both regurgitation and stenosis, complicating the clinical presentation. Tetralogy of Fallot itself is a major cause of congenital right ventricular outflow obstruction, characterized by pulmonary stenosis, ventricular septal defect, overriding aorta, and right ventricular hypertrophy. Moreover, atrioventricular septal defects, atrial septal defects, and ventricular septal defects may alter right-sided hemodynamics and mimic features of pulmonary stenosis, particularly when associated with additional lesions causing elevated right ventricular pressure. Pulmonary atresia with intact ventricular septum represents a more extreme form of right ventricular outflow obstruction and must be distinguished from critical pulmonary stenosis in the neonatal period, as management pathways differ substantially. In adult patients, the differential diagnosis broadens to include acquired causes of pulmonary outflow obstruction. Rheumatic valvular heart disease, although more commonly affecting the mitral and aortic valves, can rarely involve the pulmonary valve, resulting in commissural fusion and leaflet thickening that resemble congenital stenosis. Carcinoid heart disease represents another significant acquired etiology, in which serotonin-rich

secretory products induce fibrotic retraction of right-sided valves, producing combined stenosis and regurgitation of the pulmonary and tricuspid valves. Pulmonary embolism must also be considered, as acute or chronic thromboembolic disease can obstruct pulmonary arterial flow, leading to elevated right ventricular pressures that simulate the hemodynamic profile of pulmonary stenosis. Right heart failure of various origins may likewise present with clinical findings suggestive of pulmonary stenosis, including jugular venous distention, hepatomegaly, and exertional dyspnea, necessitating careful diagnostic imaging to differentiate intrinsic outflow obstruction from global ventricular dysfunction. Cardiac tumors, including primary cardiac sarcomas or other intracardiac masses, can obstruct the right ventricular outflow tract or pulmonary valve region, mimicking the physiology of valvular stenosis. Sarcomas in particular may infiltrate or compress the pulmonary artery, generating gradients that resemble congenital or rheumatic obstruction. Overall, distinguishing pulmonary stenosis from these congenital and acquired conditions relies on a combination of thorough clinical assessment, advanced imaging, and attention to anatomic details that define the precise site and nature of obstruction. Accurate diagnosis is essential to selecting appropriate intervention and preventing adverse outcomes across the lifespan [42][43][44].

Prognosis

The long-term outlook for individuals with pulmonary stenosis varies widely and is closely tied to the anatomical characteristics of the pulmonary valve or vessel involved and the severity of obstruction. The natural history of this condition generally follows a favorable course in the absence of critical stenosis, particularly when identified in childhood and monitored regularly. Most patients with mild or even moderate pulmonary stenosis who do not have associated congenital heart defects or genetic syndromes can expect to lead normal, active lives with minimal restrictions. These individuals often remain asymptomatic for decades, and the lesion may remain hemodynamically stable over time, requiring only periodic evaluation to monitor for progression. In many of these cases, the right ventricle adapts adequately to the modest increase in afterload, and the risk of adverse cardiac outcomes remains low [48][49]. However, the prognosis changes significantly when the degree of stenosis becomes more severe or when the anatomy is unfavorable. Patients with progressive obstruction or additional congenital anomalies face a higher likelihood of requiring intervention to prevent right ventricular failure, cyanosis, or arrhythmias. In such situations, timely recognition of worsening gradients and prompt referral for balloon valvuloplasty or surgical repair is crucial to achieving optimal outcomes. Balloon valvuloplasty, the standard of care for valvular pulmonary stenosis with a domed valve morphology, generally yields excellent results and is associated with sustained reductions in right ventricular systolic pressure and symptomatic improvement. Patients with dome-shaped pulmonary valves tend to have the most favorable prognosis, as their valve anatomy typically responds well to balloon dilation, resulting in durable enlargement of the valve orifice with low rates of restenosis. In contrast, individuals with dysplastic pulmonary valves—characterized by thickened, rigid leaflets and little commissural fusion—exhibit a less favorable response. Although balloon valvuloplasty may still be attempted in selected cases, the likelihood of achieving substantial gradient reduction is lower, and these patients often require surgical intervention. As a result, long-term outcomes in this subgroup tend to be more guarded, and recurrent interventions may be necessary [50][51].

In terms of procedural safety, balloon valvuloplasty is associated with a low incidence of major complications, and most adverse effects are transient and clinically manageable. Common minor complications include vagal responses during catheter manipulation, catheter-induced ventricular ectopy, transient right bundle branch block, and occasional episodes of high-grade atrioventricular block, which may be temporary or rarely permanent. Although infrequent, these rhythm disturbances underscore the importance of careful peri-procedural monitoring and readiness to intervene should conduction abnormalities arise [52]. A rare but serious complication following balloon valvuloplasty is the so-called “suicidal right ventricle,” a state of dynamic right ventricular outflow tract obstruction that occurs due to sudden hypercontractility of a hypertrophied right ventricle. When the obstruction becomes dynamic rather than fixed, the pressure gradient across the pulmonary valve may paradoxically fall, not because the stenosis has resolved, but because forward flow is dramatically compromised. This potentially fatal condition can lead to hemodynamic collapse if not recognized promptly. Preventive measures include the administration of beta blockers prior to intervention, which reduce hypercontractility and mitigate the risk of dynamic obstruction. Awareness and early management are essential for preventing catastrophic outcomes [51][53]. Surgical repair or revision, when necessary, generally confers excellent long-term results. Most patients experience significant improvement in exercise tolerance following surgery, reflecting improved right ventricular hemodynamics and relief of outflow obstruction. Only 15% to 20% of surgical patients require reintervention, and when reoperation is needed, it is most commonly due to the development of significant pulmonary valve regurgitation. Chronic regurgitation can impair ventricular function over time, necessitating valve replacement or additional repair. Encouragingly, supraventricular tachycardia, which may accompany longstanding pulmonary stenosis or develop postoperatively, frequently resolves after

reintervention, supporting the benefit of timely surgical follow-up in preserving long-term cardiac function [54][55].

In special populations, such as pregnant patients, the prognosis remains generally favorable. Women with mild pulmonary stenosis typically tolerate pregnancy well, with most remaining asymptomatic and requiring only routine monitoring. The high-volume circulatory demands of pregnancy may unmask exertional intolerance in some cases; however, significant maternal or fetal complications are uncommon unless the stenosis is severe. For women with severe obstruction, careful hemodynamic monitoring is essential throughout pregnancy and delivery. Although uncommon, percutaneous balloon valvuloplasty can be safely performed during pregnancy in cases of symptomatic or rapidly progressive pulmonary stenosis. This intervention may be lifesaving in selected circumstances and is generally well tolerated when performed using minimal fluoroscopy to reduce fetal radiation exposure [30]. Overall, the prognosis of pulmonary stenosis in contemporary practice is highly favorable, particularly when diagnosis occurs early and management aligns with established guidelines. Advances in catheter-based interventions, improved surgical techniques, and enhanced understanding of long-term hemodynamics have significantly reduced morbidity and mortality associated with the condition. With appropriate follow-up, most individuals—whether diagnosed during infancy, childhood, or adulthood—can anticipate excellent quality of life and near-normal life expectancy. Continuous monitoring remains vital to detect restenosis, progressive regurgitation, arrhythmias, or evolving right ventricular dysfunction, all of which are manageable when identified promptly. Through comprehensive multidisciplinary care, the long-term outlook for patients with pulmonary stenosis continues to improve, ensuring sustained functional capacity and overall favorable outcomes across the lifespan [51][52].

Complications

Complications arising from pulmonary stenosis largely reflect the chronic hemodynamic burden imposed on the right heart and the presence of underlying structural or syndromic abnormalities. Infective endocarditis is an uncommon but important complication in this population. In cases of isolated pulmonary stenosis, the overall incidence of endocarditis is low; however, certain patient groups are more vulnerable. In children, infective endocarditis is most often associated with congenitally abnormal pulmonary valves, such as those seen in Noonan syndrome, where valve dysplasia and turbulent flow create a substrate for endothelial injury and bacterial adherence [56]. In adults, the epidemiologic profile differs, and intravenous drug use becomes a leading risk factor, as contaminated injections and particulate matter can seed the right-sided valves, including the pulmonary valve, especially when pre-existing stenosis is present [56]. Beyond infective endocarditis, electrical disturbances of the heart constitute a frequent and clinically relevant complication, particularly in individuals with moderate to severe obstruction. Chronic pressure overload and right ventricular hypertrophy predispose to myocardial irritability and conduction abnormalities, increasing the risk of arrhythmias. Premature atrial contractions and premature ventricular contractions are commonly observed and may be asymptomatic or associated with palpitations, lightheadedness, or reduced exercise tolerance [57]. Ventricular couplets and more complex ventricular ectopy can also occur and may signal a higher arrhythmic burden or underlying right ventricular dysfunction [57][58]. In some patients, these arrhythmias are exacerbated following interventions such as balloon valvuloplasty or surgery, due to transient myocardial irritation, scarring, or changes in loading conditions. Over the long term, untreated or inadequately treated significant pulmonary stenosis can result in progressive right ventricular dilation, systolic dysfunction, and eventually right-sided heart failure, with manifestations such as peripheral edema, hepatomegaly, ascites, and exercise intolerance. This hemodynamic deterioration may, in turn, increase the risk of atrial arrhythmias, including atrial flutter or fibrillation. Patients with long-standing disease or those who have undergone multiple interventions may also develop significant pulmonary regurgitation, which imposes a chronic volume load on the right ventricle and further contributes to adverse remodeling. Thus, while many individuals with mild lesions enjoy an excellent prognosis, those with more advanced disease require vigilant follow-up to detect complications early, address arrhythmias, and intervene before irreversible right ventricular damage occurs [57][58].

Consultations

The evaluation and management of pulmonary stenosis frequently necessitate input from multiple subspecialists, with the specific pattern of consultation guided by the patient's age, clinical status, and associated conditions. In adults with suspected symptomatic pulmonary stenosis, early referral to an adult congenital cardiologist is essential. These clinicians possess specialized expertise in the natural history, anatomical variants, and long-term complications of congenital heart disease persisting or first detected in adulthood. They can interpret complex imaging findings, determine the need for further invasive testing, and guide decisions regarding catheter-based or surgical interventions while also addressing issues such as arrhythmia surveillance, pregnancy counseling, and long-term exercise recommendations. Pregnant patients with pulmonary stenosis represent a particularly vulnerable group in

whom multidisciplinary collaboration is critical. The hemodynamic changes of pregnancy—including increased blood volume, elevated cardiac output, and altered vascular resistance—can exacerbate right ventricular outflow obstruction and unmask previously silent disease. For this reason, a high-risk maternal-fetal medicine specialist should be involved alongside an adult congenital cardiologist. Together, they can optimize maternal and fetal monitoring, determine the timing and mode of delivery, and assess whether any cardiac intervention is necessary during pregnancy or should be deferred until the postpartum period. Anesthesia and critical care specialists may also be consulted to ensure hemodynamic stability during labor and delivery, particularly in women with severe stenosis or significant right ventricular dysfunction [56][57].

In the neonatal and pediatric setting, prompt consultation with a pediatric cardiologist is essential when pulmonary stenosis is suspected or confirmed. Newborns with critical pulmonary stenosis, in particular, require rapid assessment to determine ductal dependency, the presence of associated lesions, and the need for prostaglandin E1 infusion or urgent catheter-based intervention. Depending on the severity of the obstruction and the infant's hemodynamic status, pediatric critical care involvement may be required to manage ventilation, inotropic support, and invasive monitoring in a cardiac intensive care unit. In more complex cases, such as those associated with genetic syndromes or extracardiac anomalies, additional consultations with medical geneticists, neonatologists, and pediatric intensivists may be warranted. Across all age groups, ongoing care may also benefit from the involvement of electrophysiologists, particularly when arrhythmias complicate the clinical picture, and cardiac surgeons, when percutaneous options are limited or have failed. This structured, multidisciplinary consultation framework ensures that patients with pulmonary stenosis receive comprehensive, individualized management at each stage of life, from the neonatal period through adulthood and pregnancy [56][57].

Patient Education

Effective deterrence of complications and optimization of long-term outcomes in pulmonary stenosis hinge on comprehensive patient and family education. For pediatric patients, parents and caregivers must first understand the nature of the condition, including whether the stenosis is mild, moderate, or severe, and how this classification influences prognosis and treatment. Education should address potential therapeutic options such as balloon valvuloplasty or surgery, explaining the expected benefits, possible risks, and the likelihood of needing future interventions. Families also need guidance on recognizing warning signs of clinical deterioration, including reduced feeding or activity in infants, increased fatigue, dyspnea, syncope, or palpitations in older children. Clear instruction on the importance of routine follow-up visits, echocardiographic surveillance, and adherence to specialist recommendations is central to effective deterrence of adverse outcomes. Adult patients, particularly women of childbearing age, require detailed counseling regarding the implications of pulmonary stenosis for pregnancy and long-term health. Preconception counseling should include an assessment of stenosis severity, right ventricular function, and the potential need for intervention before pregnancy. Patients must be informed that pregnancy, with its substantial hemodynamic demands, can exacerbate symptoms or precipitate decompensation in those with significant obstruction. They should also understand that although mild pulmonary stenosis is usually well tolerated, closer monitoring is warranted in moderate or severe disease, and percutaneous intervention may occasionally be necessary during pregnancy. Additionally, educating patients about lifestyle measures, including exercise recommendations tailored to the severity of their condition, blood pressure and weight management, and prompt treatment of infections, contributes to reducing long-term risk. Genetic counseling plays an essential role for individuals and families in whom pulmonary stenosis is part of a broader congenital or syndromic context, such as Noonan, Alagille, or Williams syndromes. Counseling should address recurrence risks in future pregnancies, options for prenatal or preimplantation genetic testing, and the potential for associated cardiac and extracardiac anomalies in offspring. This information empowers families to make informed reproductive choices and prepares them for early surveillance in subsequent children. In some cases, relatives may benefit from screening echocardiography or genetic testing when familial forms of congenital heart disease are suspected. Education should also cover the principles of infective endocarditis prevention, especially in patients with prosthetic valves, prior endocarditis, or complex cyanotic congenital heart disease. Patients and parents should be taught about the importance of maintaining good dental hygiene, recognizing situations that may warrant antibiotic prophylaxis, and promptly reporting symptoms suggestive of infection. Ultimately, a well-informed patient and family are better equipped to participate actively in care, adhere to follow-up, and collaborate with healthcare providers in minimizing complications and maintaining quality of life [57][58].

Other Issues

A particularly important practical consideration in the management of pulmonary stenosis is the participation of affected individuals in athletic and competitive sports. The American Heart Association and American College of Cardiology have issued a scientific statement that provides detailed guidance for athletes with this condition. According to the 2015 AHA/ACC recommendations, athletes with mild pulmonary stenosis—defined as a Doppler-derived peak instantaneous gradient less than 40 mm

Hg with normal right ventricular function—are generally permitted to participate in all competitive sports, reflecting the low hemodynamic burden of such lesions. Annual reevaluation is advised to detect any progression in severity or changes in ventricular function. Similarly, athletes who have undergone pulmonary valvotomy or balloon valvuloplasty and have achieved adequate relief of obstruction, with residual gradients below 40 mm Hg, may also engage fully in competitive sports [59]. For athletes with more significant obstruction, stricter limitations are recommended. Those with moderate pulmonary stenosis, characterized by a Doppler-derived gradient of 40 to 60 mm Hg, or severe pulmonary stenosis, with gradients exceeding 60 mm Hg, are generally advised to limit their involvement to low-intensity class IA and IB sports. Class IA sports consist of activities with low static and low dynamic components, such as bowling, golf, and yoga, which impose relatively modest cardiovascular demands. Class IB sports involve low-static but moderately dynamic exertion, including basketball, softball, table tennis, and volleyball, where intermittent bursts of activity occur but sustained hemodynamic stress is avoided [59]. Athletes with severe pulmonary insufficiency and marked right ventricular enlargement should also restrict their participation to these lower-intensity categories, as high-intensity sports could exacerbate right ventricular dilation, arrhythmias, or heart failure [60].

In 2020, the European Association of Preventive Cardiology (EAPC) and the European Society of Cardiology's Section of Sports Cardiology and Exercise redefined the approach to exercise recommendations for adolescents and adults with congenital heart disease, including pulmonary stenosis. Rather than focusing solely on the anatomical diagnosis, the revised guidelines emphasize hemodynamic and electrophysiological parameters, integrating the patient's baseline cardiovascular status with the typical loading conditions and cardiac remodeling associated with various sports. Key parameters include ventricular function, pulmonary artery pressure, aortic dimensions, the presence or absence of arrhythmias, and arterial oxygen saturation at rest and during cardiopulmonary exercise testing [61]. When all of these parameters are within normal limits or reflect only mild abnormalities—such as mild ventricular hypertrophy or a modest pressure or volume load—patients are generally allowed to participate in all competitive sports, assuming no other contraindications. However, when any parameter is abnormal, restrictions become more specific. Patients may be advised to avoid high-endurance sports, such as long-distance running, cycling, rowing, cross-country skiing, and long-distance skating, which impose sustained high hemodynamic demands and require intensive training. Instead, they may be directed toward skill-based sports with lower cardiovascular stress, including golf, motor racing, sailing, scuba diving, table tennis, or ski jumping. For individuals with marked structural, hemodynamic, or electrophysiologic abnormalities who are symptomatic, activities are typically limited to recreational, low-intensity exercise [61]. This individualized, physiology-based approach allows clinicians to tailor recommendations to each athlete's risk profile, balancing the well-recognized benefits of physical activity against the potential cardiovascular hazards in the setting of pulmonary stenosis.

Enhancing Healthcare Team Outcomes

Optimizing outcomes for patients with pulmonary stenosis requires a coordinated, interprofessional approach that leverages the skills and expertise of a diverse healthcare team. Clinicians and advanced practitioners—including adult congenital cardiologists, pediatric cardiologists, and cardiothoracic surgeons—are central in establishing the diagnosis, characterizing the anatomy and severity of stenosis, and determining the most appropriate management strategy, whether conservative follow-up, catheter-based intervention, or surgical repair. They interpret complex imaging findings, guide the timing of intervention, and manage long-term issues such as arrhythmias, pregnancy, and exercise counseling. Nurses play a pivotal role in ensuring continuity of care across inpatient, outpatient, and home settings. They monitor vital signs, assess for changes in symptoms, recognize early warning signs of decompensation, and reinforce patient and family education. In the pediatric setting, cardiac nurses and nurse practitioners help families understand postprocedural expectations, medication regimens, and the importance of follow-up imaging. In adult care, they support patients in navigating lifestyle adjustments, adherence to medical therapy, and coordination of specialty appointments. Pediatric and adult critical care nurses are essential for managing unstable patients with critical pulmonary stenosis, administering prostaglandin infusions, titrating inotropes, and monitoring invasive hemodynamic parameters. Pharmacists contribute by ensuring the safe and effective use of medications, including agents used for heart failure, arrhythmia suppression, thromboprophylaxis, and management of comorbid conditions such as hypertension or diabetes. They counsel patients about potential drug interactions, side effects, and the importance of adherence, which is critical for therapies such as beta blockers in those at risk for dynamic right ventricular outflow obstruction after valvuloplasty. In complex cases, pharmacists also assist in optimizing antibiotic regimens for infective endocarditis prophylaxis or treatment.

Other professionals, such as genetic counselors, psychologists, dietitians, and physical therapists, further enhance care. Genetic counselors help families understand heritable aspects of pulmonary stenosis and associated syndromes, supporting informed reproductive choices.

Psychologists and social workers provide emotional and psychosocial support, addressing anxiety, depression, or adjustment issues that may arise from living with a chronic cardiac condition. Dietitians can assist in optimizing nutrition for children with growth concerns or adults with heart failure, while physical therapists and exercise specialists design safe activity plans tailored to functional status and sports participation guidelines. Effective interprofessional communication is the glue that integrates these contributions into a cohesive care plan. Regular case conferences shared electronic health records, and clear documentation of treatment goals and follow-up plans all facilitate timely information exchange. This is especially critical during transitions of care, such as from pediatric to adult congenital cardiology, or when managing pregnancy in women with pulmonary stenosis, where coordination between cardiology, obstetrics, anesthesiology, and neonatology is essential. By emphasizing patient-centered care, clearly defined roles, and open communication, the interprofessional team can reduce errors, enhance patient satisfaction, and ultimately improve clinical outcomes for individuals with pulmonary stenosis across their lifespan [61].

Conclusion:

In conclusion, pulmonary stenosis is a heterogeneous congenital condition with a generally favorable long-term prognosis when managed appropriately. The cornerstone of care is accurate diagnosis and stratification of severity using echocardiography, supplemented by advanced imaging when needed. Management is dictated by the gradient, valve morphology, and symptom status. Balloon pulmonary valvuloplasty stands as a highly effective, first-line therapy for typical valvular PS, while surgery remains crucial for dysplastic valves and complex anatomies. Critical PS in neonates represents a medical emergency, demanding prompt stabilization and intervention. Across all age groups, successful outcomes rely on a structured, multidisciplinary approach involving congenital cardiologists, interventionalists, surgeons, and specialized nurses. Lifelong surveillance is essential to monitor for potential complications such as restenosis, progressive pulmonary regurgitation, arrhythmias, and right ventricular dysfunction. Furthermore, patient-specific considerations, including genetic counseling, pregnancy planning, and guided sports participation, are integral to comprehensive care. Adherence to established guidelines and the coordinated efforts of an interprofessional healthcare team ensure that individuals with PS can achieve excellent functional capacity and quality of life from infancy through adulthood.

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