



# Cardio Policy: Intervention on Adults with Congenital Heart Defects

<b>POLICY NUMBER</b> UM CARDIO_1418	<b>SUBJECT</b> Intervention on Adults with Congenital Heart Defects		<b>DEPT/PROGRAM</b> UM Dept	<b>PAGE 1 OF 7</b>
<b>DATES COMMITTEE REVIEWED</b> 12/09/20, 06/09/21, 11/09/21, 07/13/22, 07/18/23	<b>APPROVAL DATE</b> July 18, 2023	<b>EFFECTIVE DATE</b> July 28, 2023	<b>COMMITTEE APPROVAL DATES</b> 12/09/20, 06/09/21, 11/09/21, 07/13/22, 07/18/23	
<b>PRIMARY BUSINESS OWNER:</b> UM		<b>COMMITTEE/BOARD APPROVAL</b> Utilization Management Committee		
<b>NCQA STANDARDS</b> UM 2		<b>ADDITIONAL AREAS OF IMPACT</b>		
<b>CMS REQUIREMENTS</b>	<b>STATE/FEDERAL REQUIREMENTS</b>		<b>APPLICABLE LINES OF BUSINESS</b> Commercial, Exchange, Medicaid	

## I. PURPOSE

Indications for determining medical necessity for percutaneous and surgical therapeutic interventions for adults with congenital heart disease.

## II. DEFINITIONS

Congenital heart defects (CHD) are diagnosed in 8-10 out of every 1,000 live births in the USA. Based on 2010 census data, the CDC estimates that there are currently more adults living with CHD than there are children living with CHD – a testament to improved treatments and lifespans. As such, adult cardiologists are becoming more and more likely to encounter patients with CHD who require maintenance, follow-up, and additional procedures later in life.

CHD can be broken down into five categories: shunt lesions, left-sided obstructive lesions, right-sided lesions, complex lesions, pulmonary, and coronary artery anomalies.

Shunt lesions allow for unnatural mixing of oxygenated and deoxygenated blood between the left and right circulatory systems such that a volume of blood from the higher-pressure region are transferred to that of the lower-pressure region, causing volume and pressure overload to the receiving circuit, and may result in deoxygenated blood entering the systemic circulation. They include septal defects of the atria and ventricles (ASD and VSD), atrioventricular septal defects (AVSD), anomalous

connections between the pulmonary veins and right-sided venous return to the heart (partial anomalous pulmonary venous connections (PAPVC), and patent ductus arteriosus (PDA). The purpose of correction is to maintain separation of the systemic and pulmonary circulations and to correct the resulting pressure and volume overload on the receiving circuit.

Left-sided obstructive lesions include membranous occlusions of chambers, congenital valve stenoses, and coarctation of the aorta. Correction is essential to maintain forward cardiac output. Right-sided lesions involve a combination of isolated valve stenosis and regurgitation, as well as similar issues arising from Tetralogy of Fallot (TOF). Complex lesions involve transposition of the great arteries (TGA), and abnormalities involving single ventricles with double outlets and/or double inlets, where ongoing problems usually involve valve lesions, arrhythmias, and heart failure. Anomalous coronary arteries (ACA) involve an anomalous origin of the left and right coronary arteries along with an associated abnormal anatomical pathway whose course can result in restricted blood flow and myocardial ischemia. Surgical correction is necessary to prevent ischemia, infarction, and sudden cardiac death.

The diagnoses and procedures covered here have been limited to the more common ones encountered in clinical practice. It is recognized that more complex clinical situations and interventions exist, and such requests will be determined on a case-by-case basis by clinical reviewers.

Appropriate Use Criteria (AUC score) for a service is one in which the expected incremental information, combined with clinical judgment, exceeds the expected negative consequences by a sufficiently wide margin for a specific indication that the procedure is generally considered acceptable care and a reasonable approach for the indication. The ultimate objective of AUC is to improve patient care and health outcomes in a cost-effective manner but is not intended to ignore ambiguity and nuance intrinsic to clinical decision making.

Appropriate Care- Median Score 7-9

May be Appropriate Care- Median Score 4-6

Rarely Appropriate Care- Median Score 1-3

### III. POLICY

**Indications for approving a request for medical necessity are:**

#### A. Shunt Lesions

1. ASD
  - a. In adults with isolated secundum ASD causing impaired functional capacity, right atrial and/or RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., pulmonary–systemic blood flow ratio [Qp:Qs]  $\geq 1.5:1$ ) without cyanosis at rest or during exercise, transcatheter or surgical closure to reduce RV volume and improve exercise tolerance is recommended, provided that systolic PA pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance **(AUC Score 8)**<sup>1,2,5</sup>
  - b. Adults with primum ASD, sinus venosus defect or coronary sinus defect causing impaired functional capacity, right atrial and/or RV enlargement and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ) without cyanosis at rest or during exercise, should be surgically repaired unless precluded by comorbidities, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary

vascular resistance is less than one third of the systemic vascular resistance **(AUC Score 8)**<sup>1,2,5</sup>

- c. In asymptomatic adults with isolated secundum ASD, right atrial and RV enlargement, and net left-to right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ), without cyanosis at rest or during exercise, transcatheter or surgical closure is reasonable to reduce RV volume and/or improve functional capacity, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third systemic resistance **(AUC Score 7)**<sup>1,2,5</sup>
  - d. Surgical closure of a secundum ASD in adults is reasonable when a concomitant surgical procedure is being performed and there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ) and right atrial and RV enlargement without cyanosis at rest or during exercise **(AUC Score 7)**<sup>1,2,5</sup>
  - e. Percutaneous or surgical closure may be considered for adults with ASD when net left-to-right shunt (Qp:Qs) is  $\geq 1.5:1$ , PA systolic pressure is 50% or more of systemic arterial systolic pressure, and/or pulmonary vascular resistance is greater than one third of the systemic resistance **(AUC Score 6)**<sup>1,2,5</sup>
2. VSD
- a. Adults with a VSD and evidence of left ventricular volume overload and hemodynamically significant shunts (Qp:Qs  $\geq 1.5:1$ ) should undergo VSD closure, if PA systolic pressure is less than 50% systemic and pulmonary vascular resistance is less than one third systemic **(AUC Score 8)**<sup>1,5</sup>
  - b. Surgical closure of perimembranous or supracristal VSD is reasonable in adults when there is worsening aortic regurgitation (AR) caused by VSD **(AUC Score 6)**<sup>1,5</sup>
  - c. Surgical closure of a VSD may be reasonable in adults with a history of IE caused by VSD, or in the presence of a net left-to-right shunt (Qp:Qs  $\geq 1.5:1$ ) when PA systolic pressure is 50% or more than systemic and/or pulmonary vascular resistance is greater than one third systemic **(AUC Score 5)**<sup>1,5</sup>
3. PAPVC
- a. Surgical repair is recommended for patients with partial anomalous pulmonary venous connection when functional capacity is impaired and RV enlargement is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ), PA systolic pressure is less than 50% systemic pressure and pulmonary vascular resistance is less than one third of systemic resistance **(AUC Score 8)**<sup>1,5</sup>
  - b. Repair of partial anomalous pulmonary venous connection is recommended at the time of closure of a sinus venosus defect or ASD **(AUC Score 8)**<sup>1,5</sup>
  - c. Repair of a scimitar vein (hypoplastic lung drained by an APV into systemic vein- IVC) is recommended in adults when functional capacity is impaired, evidence of RV volume overload is present, there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ), PA systolic pressure is less than 50% systemic pressure and pulmonary vascular resistance is less than one third systemic. **(AUC Score 8)**<sup>1,3,5</sup>
  - d. Surgery can be useful for right- or left-sided partial anomalous pulmonary venous connection in asymptomatic adults with RV volume overload, net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs  $\geq 1.5:1$ ), pulmonary pressures less than 50% systemic and pulmonary vascular resistance less than one third systemic **(AUC Score 6)**<sup>1,5</sup>
  - e. Surgery can be useful for repair of a scimitar vein in adults with evidence of RV volume overload, with Qp:Qs  $\geq 1.5:1$  **(AUC Score 6)**<sup>1,3,5</sup>

4. Atrioventricular Septal Defect
  - a. Surgery for severe left atrioventricular valve regurgitation is recommended per GDMT indications for mitral regurgitation **(AUC Score 7)<sup>1,5</sup>**
  - b. Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired atrioventricular septal defect is recommended when there is a net left-to-right shunt ( $Q_p:Q_s \geq 1.5:1$ ), PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic **(AUC Score 7)<sup>1,5</sup>**
  - c. Surgery for primary repair of atrioventricular septal defect or closure of residual shunts in adults with repaired septal defect is recommended when there is a net left-to-right shunt ( $Q_p:Q_s \geq 1.5:1$ ), PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic **(AUC Score 7)<sup>1,5</sup>**
5. PDA
  - a. PDA closure in adults is recommended if left atrial or LV enlargement is present and attributable to PDA with net left-to-right shunt, PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than one third systemic **(AUC Score 7)<sup>1,4,5</sup>**
  - b. PDA closure in adults may be considered in the presence of a net left-to-right shunt if PA systolic pressure is 50% or greater systemic, and/or pulmonary vascular resistance is greater than one third systemic **(AUC Score 5)<sup>1,5</sup>**

## B. Left-Sided Obstructive Lesions

1. Surgical repair is indicated for adults with cor triatriatum sinister or congenital mitral stenosis for symptoms attributable to flow obstruction **(AUC Score 8)<sup>1</sup>**
2. Surgical intervention is recommended for adults with sub-aortic stenosis with a maximum gradient of 50 mmHg or more who have symptoms attributable to the obstructive lesion, or if the gradient is less than 50 mmHg in the presence of CHF, ischemic symptoms, or LV systolic dysfunction **(AUC Score 7)<sup>1</sup>**
3. In adults with bi-leaflet aortic valve stenosis and a non-calcified valve with no more than mild AI meeting indications for intervention per GDMT (see UM CARDIO\_1095 Aortic Valve Replacement), it may be reasonable to treat with balloon valvuloplasty. **(AUC Score 5)<sup>1</sup>**  
Please refer to *UM CARDIO\_1095 Aortic Valve Replacement* or *UM CARDIO\_1295 Trans Catheter Aortic Valve Replacement* for such requests that are received for bi-leaflet AV patients.
4. Surgical repair is recommended for adults with supravalvular aortic stenosis and symptoms or decreased LV systolic function, regardless of gradient, if symptoms and LV pathology are attributable to the stenosis **(AUC Score 8)<sup>1</sup>**
5. Surgical repair or catheter-based stenting is recommended for adults with hypertension and significant (trans-obstructive gradient of 20 mmHg or more, measured by upper-lower extremity pressure differential, by echocardiography, or by cardiac catheterization) native or recurrent coarctation of the aorta **(AUC Score 8)<sup>1</sup>**

## C. Right-Sided Lesions

1. Pulmonary Valve Pathology
  - a. In adults with moderate or severe valvular pulmonary stenosis and otherwise unexplained symptoms of HF, cyanosis from interatrial right-to-left communication, and/or exercise intolerance, balloon valvuloplasty is recommended **(AUC Score 8)<sup>1,5</sup>**
  - b. If balloon valvuloplasty has failed or is not feasible, then surgical repair is recommended **(AUC Score 8)<sup>1,5</sup>**

- c. In symptomatic patients with moderate or greater PR resulting from treated isolated pulmonary stenosis, with RV dilation or RV dysfunction, pulmonary valve replacement is recommended **(AUC Score 7)<sup>1,5</sup>**
- 2. Double-Chamber Right Ventricle
  - a. Surgical repair for adults with double-chambered right ventricle and moderate or greater outflow obstruction is recommended in patients with otherwise unexplained symptoms of HF, cyanosis, or exercise limitation **(AUC Score 8)<sup>1,5</sup>**
- 3. Ebstein Anomaly
  - a. Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended when one or more of the following are present: HF symptoms, objective evidence of worsening exercise capacity, progressive RV systolic dysfunction by echocardiography or CMR **(AUC Score 8)<sup>1,5</sup>**
- 4. Tetralogy of Fallot
  - a. Pulmonary valve replacement (surgical or percutaneous) for relief of symptoms is recommended for patients with repaired TOF and moderate or greater PR with cardiovascular symptoms not otherwise explained **(AUC Score 8)<sup>1,5</sup>**
  - b. Pulmonary valve replacement (surgical or percutaneous) is reasonable for preservation of ventricular size and function in asymptomatic patients with repaired TOF and ventricular enlargement or dysfunction and moderate or greater PR **(AUC Score 6)<sup>1,5</sup>**

#### **D. Anomalous Coronary Arteries**

- 1. Surgery is recommended for anomalous origin of either the left or the right coronary artery from the opposite aortic sinus for symptoms or diagnostic evidence consistent with coronary ischemia attributable to the anomalous coronary artery **(AUC Score 8)<sup>1,5</sup>**
- 2. Surgery is reasonable for anomalous aortic origin of the left coronary artery from the right sinus in the absence of symptoms or ischemia, or if there is evidence of ventricular arrhythmia **(AUC Score 6)<sup>1,5</sup>**
- 3. Surgery is recommended for an anomalous left coronary artery that arises from the pulmonary artery **(AUC Score 8)<sup>1,5</sup>**
- 4. Surgery is recommended for an anomalous right coronary artery arising from the pulmonary artery if the patient is having symptoms that are attributed to the anomalous origin **(AUC Score 7)<sup>1,5</sup>**
- 5. Surgery is reasonable for an asymptomatic patient with an anomalous right coronary artery arising from the pulmonary artery if there is evidence of ventricular dysfunction or ischemia that is attributed to the anomalous origin **(AUC Score 6)<sup>1,5</sup>**

#### **Limitations**

- A. Closure of shunt lesions should not be performed in adults with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic, and/or a net right-to-left shunt.
- B. Requests for services that are part of a surveillance protocol for patients who are involved in a clinical trial are considered out of scope (OOS) for New Century Health and cannot be reviewed.

## **IV. PROCEDURE**

- A. In order to review a request for medical necessity, the following items must be submitted for review
  - 1. Progress notes from the cardiologist and (if indicated) cardiovascular surgeon

2. Reports from trans-thoracic and/or trans-esophageal echocardiograms, coronary/cardiac CTA, invasive cardiac catheterization, and CMR as applicable
- B. Primary codes appropriate for this service:
1. Percutaneous closure of septal defect: atrial 93580; ventricular 93581 (both include a right heart cath procedure)
  2. Surgical closure of septal defect - ASD: 33641, 33645, 33647; for VSD: 33647, 33660, 33665, 33670, 33675-7, 33681, 33684
  3. PAPVC: 33724, 33726, 33730
  4. PDA percutaneous closure: 93582; Surgical ligation: 33820, 33822, 33824; the following may be requested for surgical excision of coarctation with PDA: 33840, 33845
  5. Cor Triatriatum surgical repair: 33732
  6. Aortic valve – sub aortic membrane surgical resection: 33414-33416; Percutaneous balloon aortic valvuloplasty: 92986; Supravalvular stenosis: 33417
  7. Percutaneous repair of Coarctation of the aorta: 33881; Surgical repair (may include PDA excision): 33840, 33845
  8. Pulmonary valve percutaneous valvuloplasty: 92990; catheter-based replacement: 33477; Surgical intervention/replacement: 33470, 33471, 33474-33476, 33478
  9. Pulmonary artery percutaneous interventions: 37236, 37237; Surgical interventions: 33917, 33920, 33922, 33924-3926 Coronary artery anomalies: 33500-33507
  10. Anomalous coronary artery surgical interventions: 33500-33507

## V. APPROVAL AUTHORITY

- A. Review – Utilization Management Department
- B. Final Approval – Utilization Management Committee

## VI. ATTACHMENTS

- A. None

## VII. REFERENCES

1. Karen K. Stout et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines J of American College of Cardiology; 73 (12) Apr 2019, pp e 81–192
2. Oster M, Bhatt A, Zaragoza-Macias E, et al. Interventional therapy versus medical therapy for secundum atrial septal defect: a systematic review (part 2) for the 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol. 2018 Aug 9 [E-pub ahead of print].
3. Brink J, Yong MS, d’Udekem Y, et al. Surgery for scimitar syndrome: the Melbourne experience. Interact Cardiovasc Thorac Surg. 2015;20:31–4.
4. Gamboa R, Rios-Méndez RE, Mollón FP, et al. Percutaneous closure of patent ductus arteriosus in adults using different devices. Rev Esp Cardiol. 2010; 63:726–9.
5. ACC Appropriate Use Criteria Methodology: 2018 Update. A Report of the American College of Cardiology Appropriate Use Criteria Task Force. Hendel et al. JACC VOL. 71, NO. 8, 2018.

6. NCQA UM 2023 Standards and Elements.