

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

Indicaciones de intervención en comunicación interauricular

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms | I | B ^{2,6} |
| Device closure is the method of choice for secundum ASD closure when applicable | I | C |
| All ASDs regardless of size in patients with suspicion of paradoxical embolism (exclusion of other causes) should be considered for intervention | IIa | C |
| Patients with PVR ≥5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L-R shunt (Qp:Qs >1.5) may be considered for intervention | IIb | C |
| ASD closure must be avoided in patients with Eisenmenger physiology | III | C |

^aClass of recommendation.

^bLevel of evidence.

ASD = atrial septal defect; L-R shunt = left-to-right shunt; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; SVR = systemic vascular resistance; WU = Wood units.

Indicaciones de intervención en comunicación interventricular

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Patients with symptoms that can be attributed to L-R shunting through the (residual) VSD and who have no severe pulmonary vascular disease (see below) should undergo surgical VSD closure | I | C |
| Asymptomatic patients with evidence of LV volume overload attributable to the VSD should undergo surgical VSD closure | I | C |
| Patients with a history of IE should be considered for surgical VSD closure | IIa | C |
| Patients with VSD-associated prolapse of an aortic valve cusp causing progressive AR should be considered for surgery | IIa | C |
| Patients with VSD and PAH should be considered for surgery when there is still net L-R shunt (Qp:Qs >1.5) present and PAP or PVR are <2/3 of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) | IIa | C |
| Surgery must be avoided in Eisenmenger VSD and when exercise-induced desaturation is present | III | C |
| If the VSD is small, not subarterial, does not lead to LV volume overload or pulmonary hypertension, and if there is no history of IE, surgery should be avoided | III | C |

^aClass of recommendation.

^bLevel of evidence.

AR = aortic regurgitation; IE = infective endocarditis; L-R shunt = left-to-right shunt; LV = left ventricle; PAH = pulmonary arterial hypertension; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; VSD = ventricular septal defect.

Indicaciones de intervención en defectos del tabique atrio-ventricular

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Complete AVSD: | | |
| <ul style="list-style-type: none"> • Cardiac surgery must be avoided in patients with Eisenmenger physiology. In case of doubt, PVR testing is recommended • For indication of intervention see also VSD (Section 4.2) | III | C |
| Partial AVSD: | | |
| <ul style="list-style-type: none"> • Surgical closure should be performed in case of significant volume overload of the RV. For further details see ASD (Section 4.1) | I | C |
| AV valve regurgitation: | | |
| <ul style="list-style-type: none"> • Symptomatic patients with moderate to severe AV valve regurgitation should undergo valve surgery, preferably AV valve repair | I | C |
| <ul style="list-style-type: none"> • Asymptomatic patients with moderate or severe left-sided valve regurgitation and LVESD >45 mm and/or impaired LV function (LVEF <60%) should undergo valve surgery when other causes of LV dysfunction are excluded | I | B ³⁵ |
| <ul style="list-style-type: none"> • Surgical repair should be considered in asymptomatic patients with moderate or severe left-sided AV valve regurgitation who have signs of volume overload of the LV and a substrate of regurgitation that is very likely to be amenable for surgical repair | IIa | C |
| SubAS: | | |
| <ul style="list-style-type: none"> • See Section 4.5.3 | - | - |

^aClass of recommendation.

^bLevel of evidence.

ASD = atrial septal defect; AV = atrioventricular; AVSD = atrioventricular septal defect; LV = left ventricle; LVEF = left ventricular ejection fraction; LVESD = left ventricular end-systolic diameter; PVR = pulmonary vascular resistance; RV = right ventricle; SubAS = subaortic stenosis; VSD = ventricular septal defect.

Indicaciones de intervención en conducto arterioso persistente

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| PDA should be closed in patients with signs of LV volume overload | I | C |
| PDA should be closed in patients with PAH but PAP <2/3 of systemic pressure or PVR <2/3 of SVR | I | C |
| Device closure is the method of choice where technically suitable | I | C |
| PDA closure should be considered in patients with PAH and PAP >2/3 of systemic pressure or PVR >2/3 of SVR but still net L-R shunt (Qp:Qs >1.5) or when testing (preferably with nitric oxide) or treatment demonstrates pulmonary vascular reactivity | IIa | C |
| Device closure should be considered in small PDAs with continuous murmur (normal LV and PAP) | IIa | C |
| PDA closure should be avoided in silent duct (very small, no murmur) | III | C |
| PDA closure must be avoided in PDA Eisenmenger and patients with exercise-induced lower limb desaturation | III | C |

^aClass of recommendation.

^bLevel of evidence.

L-R shunt = left-to-right shunt; LV = left ventricle; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PDA = patent ductus arteriosus; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; SVR = systemic vascular resistance.

Indicaciones de intervención en estenosis aórtica

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Patients with severe AS and any valve-related symptoms (AP, dyspnoea, syncope) should undergo valve replacement | I | B |
| Asymptomatic patients with severe AS should undergo surgery when they develop symptoms during exercise testing | I | C |
| Regardless of symptoms, surgery should be performed when systolic LV dysfunction is present in severe AS (LVEF <50%), unless it is due to other causes | I | C |
| Regardless of symptoms, surgery should be performed when patients with severe AS undergo surgery of the ascending aorta or of another valve, or coronary artery bypass grafting | I | C |
| Regardless of symptoms, aortic surgery should be considered if the ascending aorta is >50 mm (27.5 mm ² BSA) and no other indications for cardiac surgery are present | IIa | C |
| Asymptomatic patients with severe AS should be considered for surgery when they present with a fall in blood pressure below baseline during exercise testing | IIa | C |
| Asymptomatic patients with severe AS and moderate-to-severe calcification and a rate of peak velocity progression of ≥0.3 m/s/year should be considered for surgery | IIa | C |
| Patients with moderate AS undergoing coronary artery bypass surgery or surgery of the ascending aorta or another valve should be considered for additional valve replacement | IIa | C |
| Severe AS with low gradient (<40 mmHg) and LV dysfunction with contractile reserve should be considered for surgery | IIa | C |
| Severe AS with low gradient (<40 mmHg) and LV dysfunction without contractile reserve may be considered for surgery | IIb | C |
| Asymptomatic patients with severe AS and excessive LV hypertrophy (≥15 mm), unless this is due to hypertension, may be considered for surgery | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

AP = angina pectoris; AS = aortic stenosis; BSA = body surface area; LV = left ventricle; LVEF = left ventricular ejection fraction.

Indicaciones de intervención en estenosis supravalvular aórtica

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Patients with symptoms (spontaneous or on exercise test) and mean Doppler gradient ≥ 50 mmHg should undergo surgery | I | C |
| Patients with mean Doppler gradient < 50 mmHg should undergo surgery when they have: | | |
| • symptoms attributable to obstruction (exertional dyspnoea, angina, syncope) and/or | I | C |
| • LV systolic dysfunction (without other explanation) | I | C |
| • severe LVH, attributable to obstruction (not related to hypertension) | I | C |
| • when surgery for significant CAD is required | I | C |
| Patients with mean Doppler gradient ≥ 50 mmHg ^c but without symptoms, LV systolic dysfunction, LVH, or abnormal exercise test may be considered for repair when the surgical risk is low | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

^cDoppler-derived gradients may overestimate the obstruction and may need confirmation by left heart catheterization.

CAD = coronary artery disease; LV = left ventricle; LVH = left ventricular hypertrophy.

Indicaciones de intervención en estenosis subaórtica

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥ 50 mmHg ^c or severe AR should undergo surgery | I | C |
| Asymptomatic patients should be considered for surgery when: | | |
| • LVEF is $< 50\%$ (gradient may be < 50 mmHg due to low flow) | IIa | C |
| • AR is severe and LVESD > 50 mm (or 25 mm/m ² BSA) and/or EF $< 50\%$ ^d | IIa | C |
| • mean Doppler gradient is ≥ 50 mmHg ^c and LVH marked | IIa | C |
| • mean Doppler gradient is ≥ 50 mmHg ^c and blood pressure response is abnormal on exercise testing | IIa | C |
| Asymptomatic patients may be considered for surgery when: | | |
| • mean Doppler gradient is ≥ 50 mmHg ^c , LV normal, exercise testing normal, and surgical risk low | IIb | C |
| • progression of AR is documented and AR becomes more than mild (to prevent further progression) | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

^cDoppler-derived gradients may overestimate the obstruction and may need confirmation by cardiac catheterization.

^dSee ESC guidelines on the management of valvular heart disease.³⁵

AR = aortic regurgitation; BSA = body surface area; EF = ejection fraction; LV = left ventricle; LVEF = left ventricular ejection fraction; LVESD = left ventricular end systolic diameter; LVH = left ventricular hypertrophy.

Indicaciones de intervención en coartación aórtica

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| All patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention | I | C |
| Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention | IIa | C |
| Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

Indicación de cirugía aórtica en el síndrome de Marfan

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Patients should undergo surgery when aortic root maximal diameter is: | | |
| • >50 mm | I | C ^c |
| • 46–50 mm with | I | C |
| - family history of dissection or | I | C |
| - progressive dilation >2 mm/year as confirmed by repeated measurement or | I | C |
| - severe AR or MR or | I | C |
| - desire of pregnancy | I | C |
| • Patients should be considered for surgery when other parts of the aorta >50 mm or dilation is progressive | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

^cESC guidelines for valvular heart disease are slightly more strict, recommending only one diameter (45 mm) regardless of other findings.

AR = aortic regurgitation; MR = mitral regurgitation.

Indicaciones de intervención en obstrucción del tracto de salida del ventrículo derecho

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is >64 mmHg (peak velocity >4m/s), provided that RV function is normal and no valve substitute is required | I | C |
| In valvular PS, balloon valvotomy should be the intervention of choice | I | C |
| In asymptomatic patients in whom balloon valvotomy is ineffective and surgical valve replacement is the only option, surgery should be performed in the presence of a systolic RVP >80 mmHg (TR velocity >4.3 m/s) | I | C |
| Intervention in patients with gradient <64 mmHg should be considered in the presence of: <ul style="list-style-type: none"> * symptoms related to PS or, * decreased RV function or, * double-chambered RV (which is usually progressive) or, * important arrhythmias or, * right-to-left shunting via an ASD or VSD. | IIa | C |
| Peripheral PS, regardless of symptoms, should be considered for repair if >50% diameter narrowing and RV systolic pressure >50 mmHg and/or lung perfusion abnormalities are present | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

ASD = atrial septal defect; PS = pulmonary stenosis; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; RVP = right ventricular pressure; TR = tricuspid regurgitation; VSD = ventricular septal defect.

Indicaciones de intervención en anomalía de Ebstein

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Indications for surgery | | |
| • Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class >II or arrhythmias) or deteriorating exercise capacity measured by CPET | I | C |
| • If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair | I | C |
| • Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray | IIa | C |
| Indications for catheter intervention | | |
| • Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery | I | C |
| • In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered | IIa | C |
| • If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text) | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

ASD = atrial septal defect; CPET = cardiopulmonary exercise testing; NYHA = New York Heart Association; PFO = patent foramen ovale; RV = right ventricle; TR = tricuspid regurgitation; VSD = ventricular septal defect.

Indicaciones de intervención en Tetralogía de Fallot corregida

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Aortic valve replacement should be performed in patients with severe AR with symptoms or signs of LV dysfunction | I | C |
| PVRep should be performed in symptomatic patients with severe PR and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5 m/s) | I | C |
| PVRep should be considered in asymptomatic patients with severe PR and/or PS when at least one of the following criteria is present: <ul style="list-style-type: none"> - Decrease in objective exercise capacity - Progressive RV dilation - Progressive RV systolic dysfunction - Progressive TR (at least moderate) - RVOTO with RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) - Sustained atrial/ventricular arrhythmias | IIa | C |
| VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

AR = aortic regurgitation; LV = left ventricle; PR = pulmonary regurgitation; PVRep = pulmonary valve replacement; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; TR = tricuspid regurgitation; VSD = ventricular septal defect.

Indicaciones de intervención en transposición de los grandes vasos post switch atrial

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Indications for surgical intervention | | |
| Valve repair or replacement should be performed in patients with severe symptomatic systemic (tricuspid) AV valve regurgitation without significant ventricular dysfunction (RVEF \geq 45%) | I | C |
| Significant systemic ventricular dysfunction, with or without TR, should be treated conservatively or eventually with cardiac transplantation | I | C |
| LVOTO if symptomatic or if LV function deteriorates should be treated surgically | I | C |
| In symptomatic pulmonary venous obstruction surgical repair (catheter intervention rarely possible) should be performed | I | C |
| Symptomatic patients with baffle stenosis not amenable for catheter intervention should be treated surgically | I | C |
| Symptomatic patients with baffle leaks not amenable for stenting should be treated surgically | I | C |
| Valve repair or replacement should be considered for severe asymptomatic systemic (tricuspid) AV valve regurgitation without significant ventricular dysfunction (RVEF \geq 45%) | IIa | C |
| Pulmonary artery banding in adult patients, to create septal shift, or as left ventricular training with subsequent arterial switch, is currently experimental and should be avoided | III | C |
| Indications for catheter intervention | | |
| Stenting should be performed in symptomatic patients with baffle stenosis | I | C |
| Stenting (covered) or device closure should be performed in symptomatic patients with baffle leaks and substantial cyanosis at rest or during exercise | I | C |
| Stenting (covered) or device closure should be performed in patients with baffle leaks and symptoms due to L-R shunt | I | C |
| Stenting (covered) or device closure should be considered in asymptomatic patients with baffle leaks with substantial ventricular volume overload due to L-R shunt | IIa | C |
| Stenting should be considered in asymptomatic patients with baffle stenosis who require a PM | IIa | C |
| Stenting may be considered in other asymptomatic patients with baffle stenosis | IIIb | C |

^aClass of recommendation.

^bLevel of evidence.

AV = atrioventricular; L-R shunt = left-to-right shunt; LV = left ventricle; LVOTO = left ventricular outflow tract obstruction; PM = pacemaker; RVEF = right ventricular ejection fraction; TR = tricuspid regurgitation.

Indicaciones de intervención en transposición de los grandes vasos luego del switch arterial

| Indications | Class ^a | Level ^b |
|---|--------------------|--------------------|
| Stenting or surgery (depending on substrate) should be performed for coronary artery stenosis causing ischaemia | I | C |
| Surgical repair of RVOTO should be performed in symptomatic patients with RV systolic pressure >60 mmHg (TR velocity >3.5 m/s) | I | C |
| Surgical repair of RVOTO should be performed regardless of symptoms when RV dysfunction develops (RVP may then be lower) | I | C |
| Surgical repair should be considered in asymptomatic patients with RVOTO and systolic RVP >80 mmHg (TR velocity >4.3 m/s) | IIa | C |
| Aortic root surgery should be considered when the (neo-)aortic root is >55 mm, providing average adult stature (for aortic valve replacement for severe AR see guidelines for AR ²⁵) | IIa | C |
| Stenting or surgery (depending on substrate) should be considered for peripheral PS, regardless of symptoms, if >50% diameter narrowing and RV systolic pressure >50 mmHg and/or lung perfusion abnormalities are present | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

AR = aortic regurgitation; AV = atrioventricular; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; RVP = right ventricular pressure; TR = tricuspid regurgitation.

Indicaciones de intervención en transposición de los grandes vasos corregida

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Systemic AV valve (tricuspid valve) surgery for severe regurgitation should be considered before systemic (subaortic) ventricular function deteriorates (before RVEF <45%) | IIa | C |
| Anatomic repair (atrial switch + arterial switch or Rastelli when feasible in case of non-restrictive VSD) may be considered when LV is functioning at systemic pressure | IIb | C |

^aClass of recommendation.

^bLevel of evidence.

AV = atrioventricular; LV = left ventricle; RVEF = right ventricular ejection fraction; VSD = ventricular septal defect.

Ventrículo único: consideraciones especiales e indicaciones de intervención

| Special considerations and indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Only well-selected patients after careful evaluation [low pulmonary vascular resistances, adequate function of the AV valve(s), preserved ventricular function] should be considered candidates for a Fontan operation | IIa | C |
| Patients with increased pulmonary blood flow—unlikely at adult age—should be considered for PA banding or tightening of a previously placed band | IIa | C |
| Patients with severe cyanosis, with decreased pulmonary blood flow without elevated PVR, should be considered for a bidirectional Glenn shunt | IIa | C |
| Heart transplantation and heart–lung transplantation should be considered when there is no conventional surgical option in patients with poor clinical status | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

AV = atrioventricular; PA = pulmonary artery; PVR = pulmonary vascular resistance.

Indicaciones de intervención en pacientes con conductos de ventrículo derecho a arteria pulmonar

| Indications | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Symptomatic patients with RV systolic pressure >60 mmHg (TR velocity >3.5 m/s; may be lower in case of reduced flow) and/or moderate/severe PR should undergo surgery | I | C |
| Asymptomatic patients with severe RVOTO and/or severe PR should be considered for surgery when at least one of the following criteria is present: <ul style="list-style-type: none"> • Decrease in exercise capacity (CPET) • Progressive RV dilation • Progressive RV systolic dysfunction • Progressive TR (at least moderate) • RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) • Sustained atrial/ventricular arrhythmias | IIa | C |

^aClass of recommendation.

^bLevel of evidence.

CPET = cardiopulmonary exercise testing; PR = pulmonary regurgitation; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; TR = tricuspid regurgitation.

Recomendaciones para tratamiento de hipertensión pulmonar en cardiopatías congénitas

| Recommendations | Class ^a | Level ^b |
|--|--------------------|--------------------|
| Targeted PAH therapy in CHD should only be performed in specialized centres | I | C |
| The ERA bosentan should be initiated in WHO-FC III ^c patients with Eisenmenger syndrome | I | B |
| Other ERAs, phosphodiesterase type-5 inhibitors, and prostanooids should be considered in WHO-FC III ^c patients with Eisenmenger syndrome | IIa | C |
| Combination therapy may be considered in WHO-FC III ^c patients with Eisenmenger syndrome | IIb | C |
| The use of calcium channel blockers should be avoided in patients with Eisenmenger syndrome | III | C |

^aClass of recommendation.
^bLevel of evidence.
^cAlthough recent data support the use of ERAs such as bosentan also in WHO-FC II in patients with idiopathic PAH and PAH associated with connective tissue diseases, such data are currently not available for Eisenmenger patients. Because of marked differences in the natural history between these groups, the results cannot simply be applied to congenital patients, and further studies are required before recommendations.
 CHD = congenital heart disease; ERA = endothelin receptor antagonist; PAH = pulmonary arterial hypertension; WHO-FC = World Health Organization-functional class.

Estrategias para reducción de riesgo en pacientes con cardiopatías congénitas cianóticas

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|---|
| Prophylactic measures are the mainstay of care to avoid complications. The following exposures/activities should be avoided: |
| • Pregnancy |
| • Iron deficiency and anaemia (no routine, inappropriate phlebotomies to maintain a pre-determined haemoglobin) |
| • Dehydration |
| • Infectious disease: annual influenza vaccination, pneumovax (every 5 years) |
| • Cigarette smoking, recreational drug abuse including alcohol |
| • Transvenous PM/ICD leads |
| • Strenuous exercise |
| • Acute exposure to heat (sauna, hot tub/shower) |
| Other risk reduction strategies include: |
| • Use of an air filter in an intravenous line to prevent air embolism |
| • Consultation of a GUCH cardiologist before administration of any agent and performance of any surgical/interventional procedure |
| • Prompt therapy of upper respiratory tract infections |
| • Cautious use or avoidance of agents that impair renal function |
| • Contraceptive advice |

GUCH = grown-up congenital heart disease; ICD = implantable cardioverter defibrillator; PM = pacemaker.

