Clinical Guidelines

ATRIAL SEPTAL DEFECT (ASD, INCLUDING SECUNDUM, SINUS VENOSUS AND CORONARY SINUS DEFECTS. PRIMUM ASD IS PART OF AVSD SPECTRUM)

SETTING South West England

GUIDELINE FOR Cardiology teams in South West England and South Wales hospitals

PATIENT GROUP Adult patients with congenital heart disease

GUIDANCE

Follow-up: discharge if secundum ASD surgically repaired with no sequelae
annually if pre-closure (by any method) PH, atrial arrhythmias, RV/LV dysfunction, any other lesions
4-yearly if - unoperated and no current indication for closure
- repaired sinus venosus ASD (SVASD)
[device closure patients seen in device clinic (see Device Clinic Guideline)]
See separate SOP for stent repair of SVASD

Associated lesions: partial anomalous venous drainage of right pulmonary veins (common with SVASD, occasionally with secundum ASDs)
persistent left SVC to coronary sinus with coronary sinus defects
mitral valve prolapse
pulmonary stenosis

Inheritance: risk of CHD if mother affected - 5%
associated with Holt-Oram syndrome (autosomal dominant)
autosomal dominant ASD with AV conduction defect (mutations in NKX2.5 gene)

Long-term complications: arrhythmias (late AF in up to 1/3, especially if > 40 years and/or if atrial arrhythmias pre-operatively)
residual/recurrent ASD uncommon
SVC/pulmonary vein stenosis may occur after sinus venosus ASD closure
pericardial effusion and tamponade can occur in weeks after surgical closure ASD (may present with fever, fatigue, vomiting, chest pain, or abdominal pain –immediate echo)
if device closure, migration or erosion can occur (see Device Clinic Guideline)

At each visit:

History: symptoms increase with age as shunt increases due to decreasing LV compliance
exertional dyspnoea, fatigue, palpitations
frequent chest infections
right heart failure
symptomatic supraventricular arrhythmias (AF, flutter, or sick sinus syndrome)
may have had paradoxical embolism resulting in TIA/stroke

chest pain or syncope if device erosion (urgent echo)

Exam: precordial lift
systolic pulmonary flow murmur
fixed splitting of S2 (not invariable)
diastolic flow rumble across the tricuspid valve with large shunts

ECG: check rhythm
right axis deviation if right heart dilatation
partial right bundle branch block
if first degree AV block consider autosomal dominant type ASD
atrial flutter/AF not uncommon as older
abnormal P-wave axis (in sinus venosus ASD)

Echo: visualise shunt using colour Doppler
dilated RA and RV (RV volume overload best characterizes the haemodynamic relevance of the defect and is better than shunt ratio)
RV function
tricuspid regurgitation
paradoxical motion of interventricular septum
estimate RVSP from TR
if device closure, check position and for residual shunt, thrombus formation or pericardial effusion

Further Investigations:

CXR: increased pulmonary vascular markings if significant left-to-right shunt
RV and RA enlargement
prominent MPA

CPET: indicated in patients with PAH to assess for desaturation

Holter: if symptomatic only

TOE: to assess size, location and suitability for percutaneous closure
to identify pulmonary veins

Catheter: required to calculate PVR if non-invasive signs of PH

EP study: for refractory atrial arrhythmias

MRI: to assess RV volume and function and shunt size
identification of pulmonary veins
identification of sinus venosus defects

Drugs: none

Pregnancy: low risk, even if unrepaired
risk of paradoxical embolism is increased, especially post-partum, prophylactic LMWH used

Contraception: avoid combined pill in unrepaired

Endocarditis: antibiotic prophylaxis not indicated, apart from for first 6 months after surgical repair or device closure

Exercise: no restriction in asymptomatic patients before or after intervention if no PH. low-intensity only if PH.

Discuss if:

- Unrepaired with no PH and evidence of RV volume overload (regardless of symptoms), paradoxical embolism, or platypnea –orthodeoxia (coronary sinus defectsneed to be repaired surgically). Sinus venosus defects may be assessed for stent repair.
- Unrepaired with evidence of PH (closure may be considered if net left-to-right shunt, Qp:QS >1.5, where PVR is 3-5WU. If PVR >5WU, fenestrated closure may be considered after targeted PAH therapy).
- New onset AF/flutter

Appendix 1 – Evidence of Learning from Incidents

The following table sets out any incidents/ cases which informed either the creation of this document or from which changes to the existing version have been made.

Incidents	Summary of Learning
n/a	

Table A

REFERENCES	Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, Lung B, Kluin J, Lang IM, Meijboom F, Moons P. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2020. Stout et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. Journal of the American College of Cardiology Aug 2018, 25255; DOI: 10.1016/j.jacc.2018.08.1029 Rychik J, Atz AM, Celermajer DS, Deal BJ, Gatzoulis MA, Gewillig MH, Hsia TY, Hsu DT, Kovacs AH, McCrindle BW, Newburger JW, Pike NA, Rodefeld M, Rosenthal DN, Schumacher KR, Marino BS, Stout K, Veldtman G, Younoszai AK, d'Udekem Y; American Heart Association Council on Cardiovascular Disease in the Young and Council on Cardiovascular and Stroke Nursing. Evaluation and Management of the Child and Adult With Fontan Circulation: A Scientific Statement From the American Heart Association. Circulation. 2019 Jul 1. Greenway SC, Crossland DS, Hudson M, Martin SR, Myers RP, Prieur T, et al. Fontan-associated liver disease: Implications for heart transplantation. The Journal of Heart and Lung Transplantation 2016;35:26-33.
RELATED DOCUMENTS AND PAGES	Regional Referral Guidance for Adult Patients with Congenital Heart Disease RegionalReferralGuidanceAdultPatientsWithCongenita-3.pdf Regional Referral Pathway for Cardiac Disease in Pregnancy ClinicalGuidelineForCardiacDiseasePreExistingOrPre-1.pdf

AUTHORISING BODY	Cardiac Executive Group, Bristol Heart Institute
SAFETY	None
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AUDIT REQUIREMENTS	Adherence to guideline will be audited periodically as part of ACHD departmental audit

Plan Elements	**Plan Details**
The Dissemination Lead is:	Dr Stephanie Curtis
Is this document: A – replacing the same titled, expired SOP, B – replacing an alternative SOP, C – a new SOP:	A
If answer above is B: Alternative documentation this SOP will replace (if applicable):	
This document is to be disseminated to:	South West and South Wales Congenital Heart Network
Method of dissemination:	Email
Is Training required:	No

Document Change Control

Date of Version	Version Number	Lead for Revisions	Type of Revision	Description of Revision
Jan 2021	2	Consultant Cardiologist	Minor	Updated contacts and related documents. Inheritance risk changed to 5% Follow up changed to: - discharge if secundum ASD surgically repaired with no sequelae - annually if pre-closure (by any method) PH,

				atrial arrhythmias, RV/LV dysfunction, any other lesions - 4-yearly if unoperated and no current indication for closure repaired sinus venosus ASD (SVASD) [Device closure patients seen in device clinic (see Device Clinic Guideline)] See separate SOP for stent repair of SVASD Under pregnancy “prophylactic LMWH used” added.