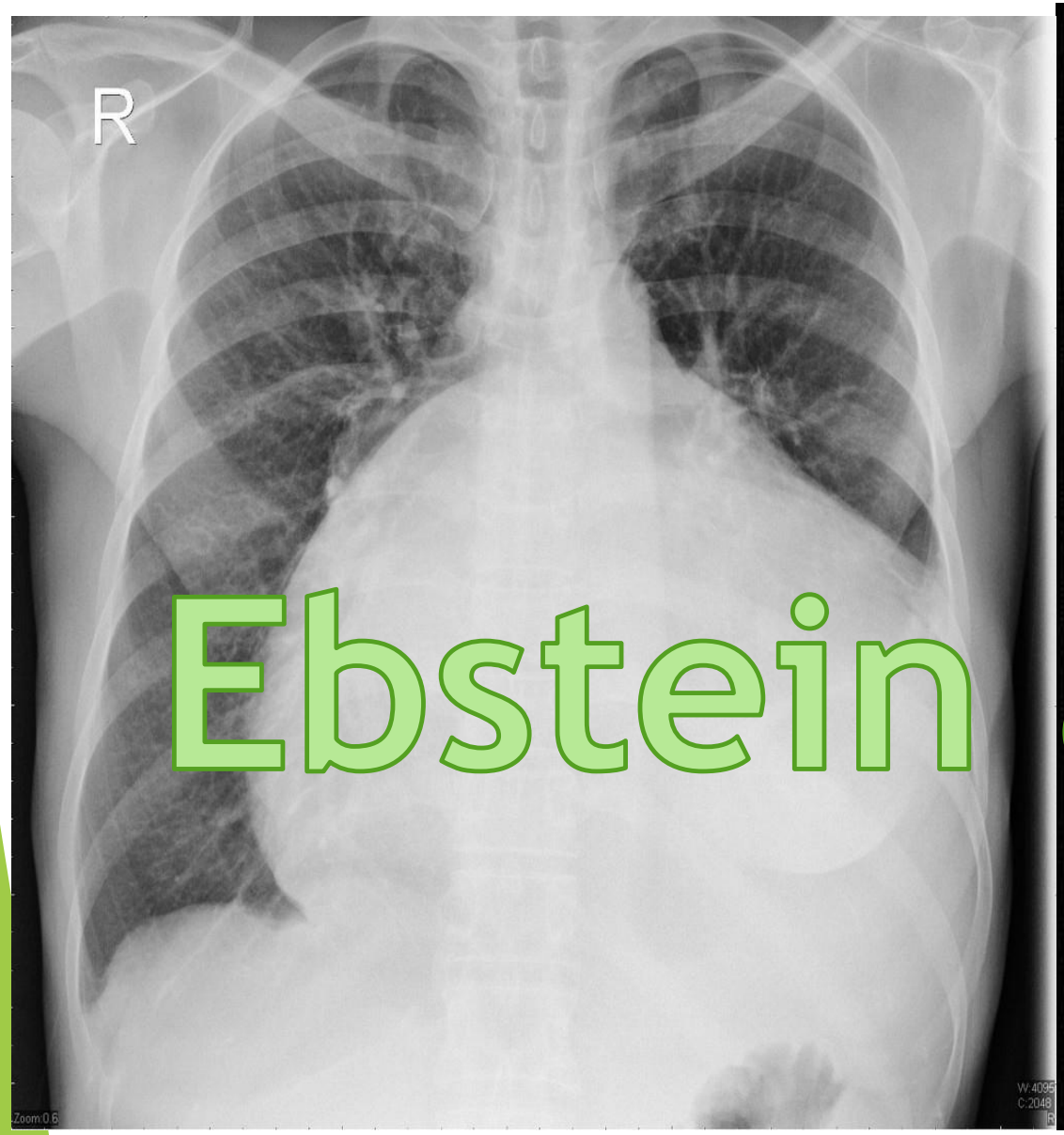
اعداد : د محمد المجبل

حالة سريرية

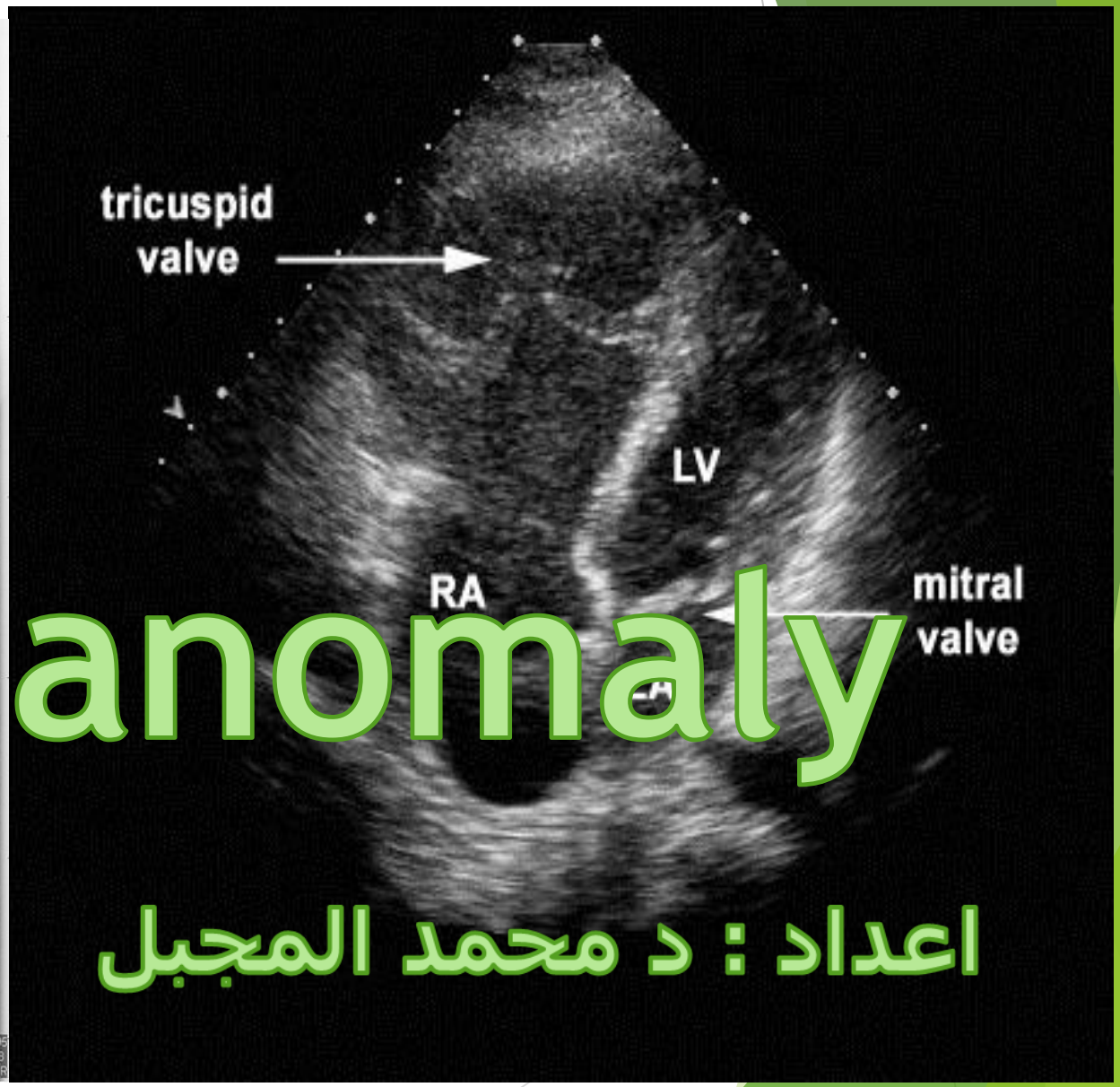
خفقان متكرر

انتانات صدرية متكررة

كورونا



Ebstein anomaly



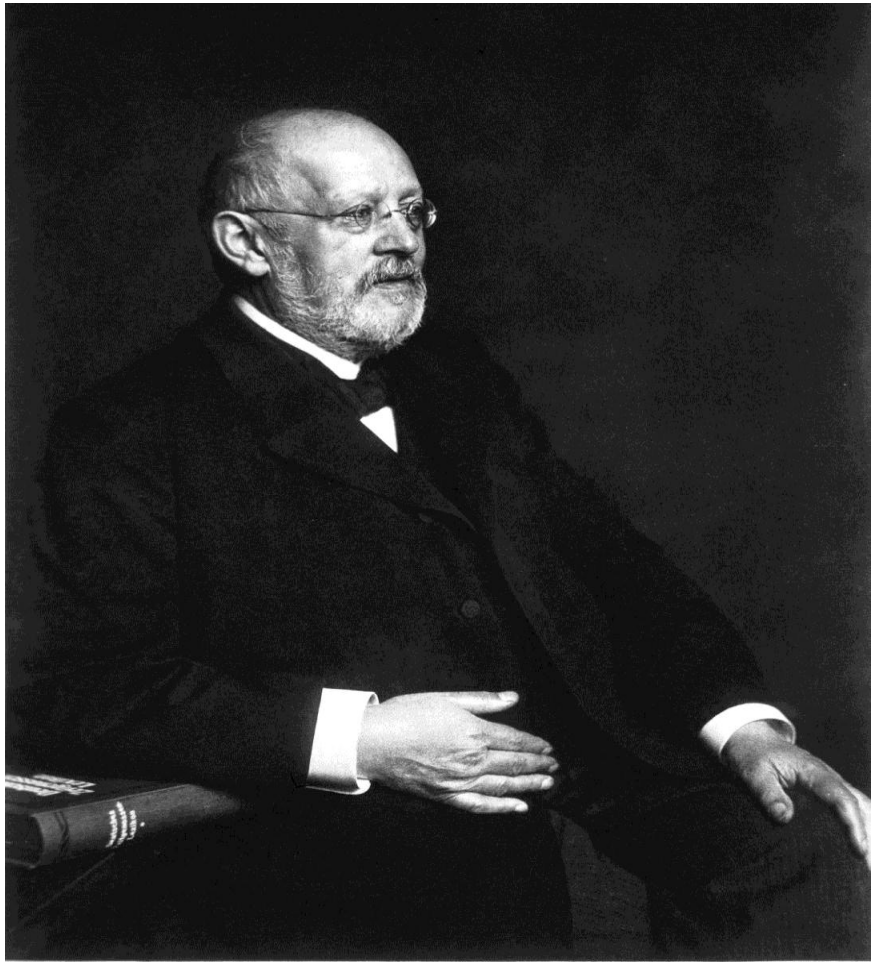
اعداد : د محمد المجبل

ESC

European Society of Cardiology

2020 ESC Guidelines for the management of adult congenital heart disease

يشكل أقل من ١ % من التشوهات الخلقية



أول من وصفه ١٨٦٦
Wilhelm Ebstein

abnormally formed and apically displaced leaflets of the TV

septal and posterior leaflets are displaced towards the RV apex

right heart consists of a morphological RA, an atrialized \ portion of the RV

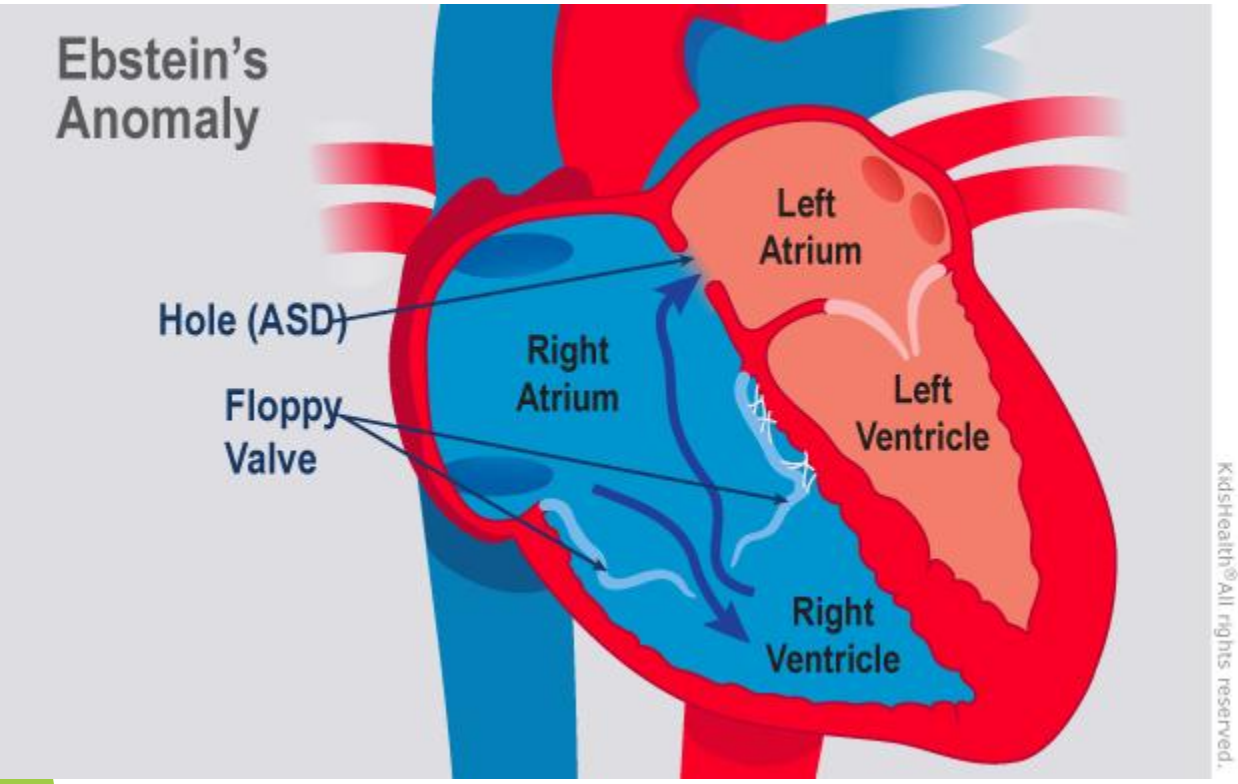
TV is often regurgitant

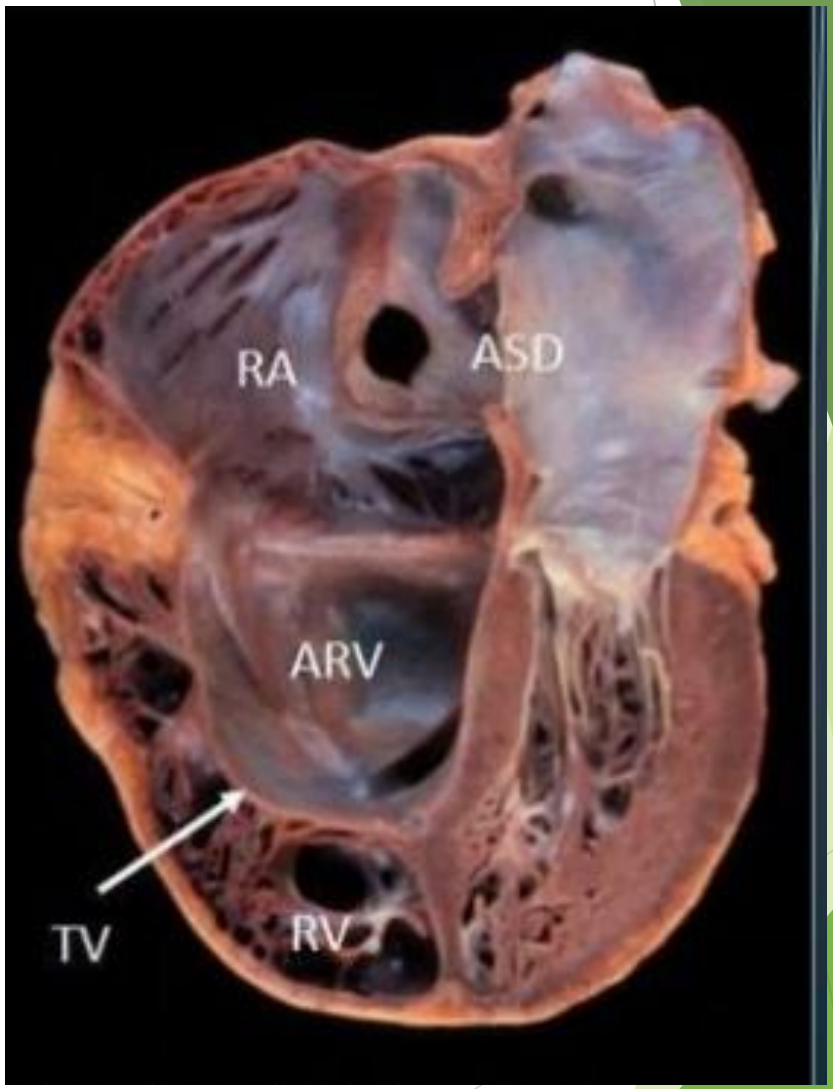
secundum ASD or patent foramen ovale (PFO)

accessory pathways, including Mahaim-type pathways

Associated defects

- Commonly associated with:
  - ASD or PFO (90%)
  - VSD, AV canal defect
  - Pulmonary stenosis/atresia (20-25%)
  - Wolff-Parkinson-White
- Syndromes:
  - Down, Marfan, Noonan


Ebstein's Anomaly
Hole (ASD)
Floppy Valve
Right Atrium
Left Atrium
Left Ventricle
Right Ventricle
KidsHealth® All rights reserved.


RA
ASD
ARV
TV
RV

Clinical presentation and natural history

atrioventricular reentrant tachycardia (AVRT)

high-grade TR, RV dysfunction, RV failure

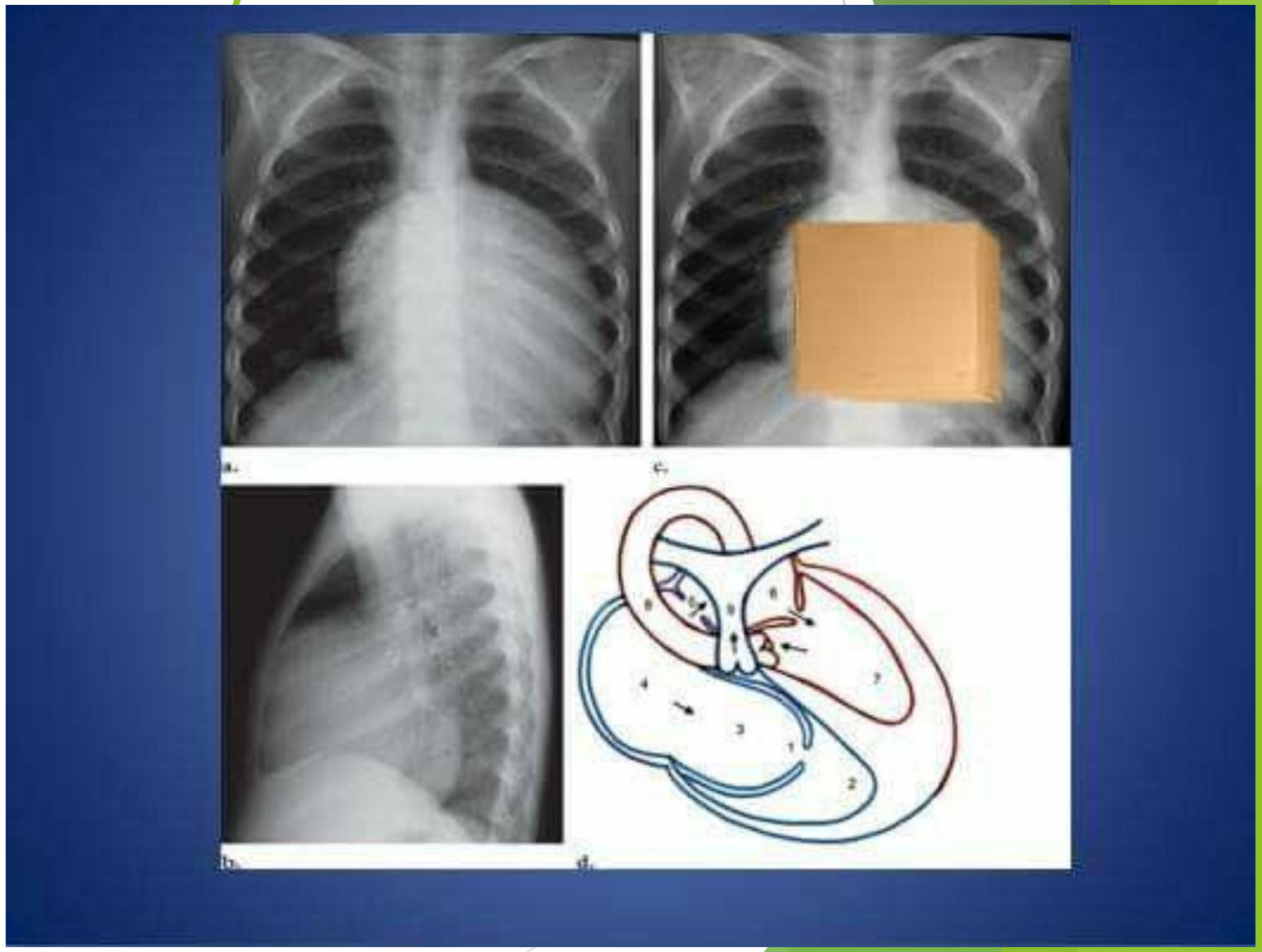
Liver cirrhosis, cerebral abscesses, paradoxical embolism , pulmonary embolism, tachyarrhythmias

dyspnoea, fatigue, poor exercise tolerance, chest pain, and peripheral and/or central cyanosis.

- Fetal life:
  - Diagnosed incidentally by echocardiography.
- Neonatal life and infancy:
  - Cyanosis and/or severe heart failure
  - Improve as pulmonary vascular resistance decreases.
- Adult life:
  - Fatigue, exertional dyspnea, cyanosis, tricus pid regurgitation and/or right heart failure, and

Chest X-ray

- squared-off cardiac contour
- pulmonary oligemia



Surgical/catheter interventional treatment

If there is an indication for TV surgery, ASD/ PFO closure is recommended at the time of valve repair if it is expected to be haemodynamically tolerated.	I	C
Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function.	IIa	C

Follow-up recommendations

Regular follow-up (at least yearly) is required in all patients in specialized ACHD centres

Typical post-operative residual anomalies to look for are persisting or new TR

Reintervention may become necessary for recurrent TR and failure of prosthetic valves.

Exercise/sports

patients without residual anomalies can usually lead normally active lives without restriction

Pregnancy

asymptomatic females with good ventricular function may tolerate pregnancy well

There is a certain risk of RV failure, arrhythmia, and paradoxical embolism

Pregnancy will be of higher risk in the presence of significant cyanosis, serious arrhythmia, and right heart failure

IE prophylaxis:

recommended only for high-risk patients