International Journal of Obstetric Anesthesia (2010) **19**, 306–312
0959-289X/$ - see front matter 
doi:10.1016/j.ijoa.2009.10.009

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REVIEW ARTICLE

# A functional understanding of moderate to complex congenital heart disease and the impact of pregnancy. Part II: Tetralogy of Fallot, Eisenmenger's syndrome and the Fontan operation

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## Introduction

Since the introduction of cardiopulmonary bypass procedures, the survival of babies with moderate to complex congenital heart disease (CHD) has become the norm. More than 85% survive to adulthood.[1] As a result a new cohort of women wishes to risk the cardiovascular challenges of pregnancy and childbirth.

In this article we review three important congenital heart disease circulations, Tetralogy of Fallot, Eisenmenger's syndrome and the Fontan circulation. These articles concentrate on haemodynamic variables rather than anatomic details. We feel an understanding of these factors will improve communication between anaesthetists, surgeons and cardiologists, when managing pregnancy, anaesthesia, surgery and delivery in these conditions.

Accepted October 2009
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## Fallot's tetralogy

First described in 1888 by the French physician Etienne-Louis Arthur Fallot,[2] tetralogy of Fallot is a relatively common cyanotic congenital heart disease with an incidence of 10% of all reported congenital heart diseases.[3] It is characterised by four components (Fig. 1a):

1. Large ventricular septal defect (VSD).
2. Right ventricular outflow tract obstruction. This is variable, almost universally at a subvalvular level, even at the level of the pulmonary artery, but often associated with an abnormality in the pulmonary valve itself. This obstruction causes a right to left

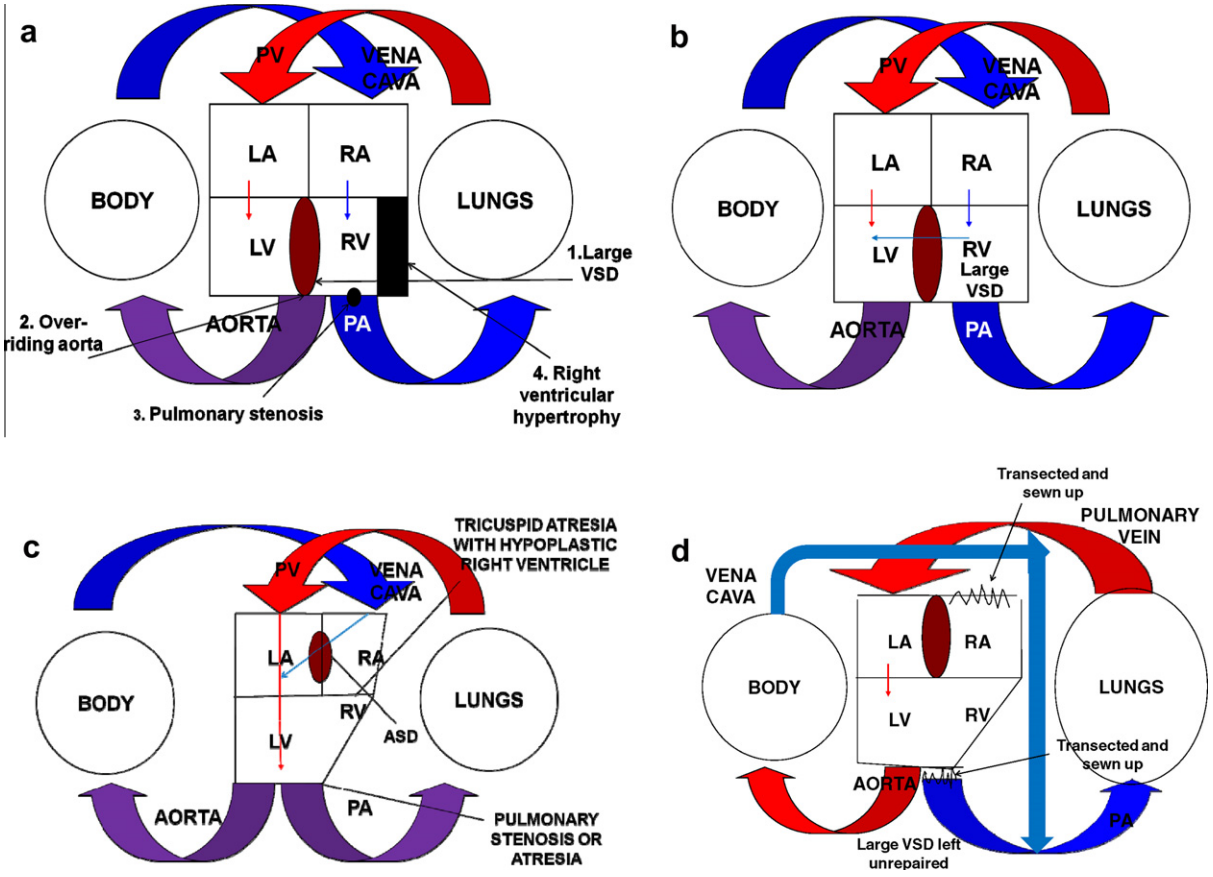


**Fig. 1** (a) Fallot's Tetralogy. The purple colour represents mixed oxygenated and de-oxygenated blood. (b) Eisenmenger's circulation. The purple colour represents mixed oxygenated and de-oxygenated blood. (c) Univentricular circulation suitable for the Fontan operation. The purple colour represents mixed oxygenated and de-oxygenated blood. (d) Total cavopulmonary connection (TCPC), the modern Fontan circulation. LA = left atrium, LV = left ventricle, RA = right atrium, ASD = atrial septal defect, RV = right ventricle, VSD = ventricular septal defect, PA = pulmonary artery, PV = pulmonary veins. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

shunt, a reduction in blood flow to the pulmonary artery and subsequent cyanosis.
3. Overriding aorta.
4. Right ventricular hypertrophy. Progressive right ventricular outflow tract obstruction results in right ventricular hypertrophy which adds to the obstruction and worsens the condition.[4]

Prognosis for patients who do not undergo surgical repair is poor; the most common causes of death are pulmonary haemorrhage, brain abscess and thromboembolic complications. In 1945 the first surgical treatment for tetralogy of Fallot was performed by Blalock and Taussig.[5] This was followed by the first intracardiac repair in 1954 and one year later the first repair using a pump oxygenator by Kirklin.[6] Palliation of tetralogy of Fallot with systemic to pulmonary shunts such as the Blalock shunt has been the accepted standard for symptomatic neonates and infants.[7] The Blalock shunt procedure involves fashioning a conduit from the subclavian to the pulmonary artery to increase blood flow to the lungs and bypass pulmonary stenosis. Indications for palliative treatment include cyanotic spells or persistent profound arterial desaturation, very young age, or unfavourable pulmonary arterial anatomy.[8]

## Surgical repair

Primary repair of tetralogy of Fallot is routinely performed in stable patients aged 6 months or older with suitable pulmonary artery anatomy. The best timing for the repair remains controversial, varying between 6 months and 2 years, although currently many surgeons are considering primary neonatal repair.[9] Repair involves relieving the right ventricular outflow obstruction and closure of the VSD.

Several complications may follow surgical repair. Up to 80% of patients develop a degree of pulmonary regurgitation.[10] This is made worse by factors that elevate pulmonary artery pressure. Although well tolerated in early life, pulmonary regurgitation may lead to right ventricular dilatation and failure, reduced exercise