Fetal Cardiology

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Editors

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A Practical Approach to Diagnosis and Management

Springer

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Foreword

This book represents a major collaborative effort between experts in fetal cardiology, fetal medicine and genetics from recognised centres of excellence in the UK. It provides a plethora of images and videos and extensive description of the essential views for examination of the anatomy and function of the fetal heart and features of the major types of congenital heart defects.

In the case of heart defects, the reader will gain a clear understanding of the diagnosis and the need to search for other abnormalities by detailed ultrasound examination. Invasive testing, examination of cell-free DNA in maternal blood, parental counselling and postnatal care are also discussed. To date, fetal cardiac imaging has been almost exclusively the preserve of ultrasound. This book includes the latest advances in fetal cardiac MRI to investigate anatomic abnormalities and assessment of blood flow.

The book provides essential reading for experts in fetal medicine and paediatric cardiology but also for obstetricians, sonographers and midwives because fetal echocardiography and the diagnosis and management of fetal heart abnormalities constitute an integral part of prenatal care.

Kypros Nicolaides

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Vita Zidere

I acknowledge the support of my wife Vanda, and of my children Hamish, Maddy and Angus, whilst putting together this book.

Owen I. Miller

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Abbreviations

A	Atrium
Ao	Aorta
ALSA	Aberrant left subclavian artery
ARSA	Aberrant right subclavian artery
AVSD	Atrioventricular septal defect
bpm	Beats per minute
CAT	Common arterial trunk
CHD	Congenital heart disease
CoA	Coarctation of the aorta
CPAM	Congenital pulmonary airway malformation
CTR	Cardio-thoracic ratio
D	Duct (arterial)
DILV	Double inlet left ventricle
DORV	Double outlet right ventricle
HLH	Hypoplastic left heart
IA	Innominate artery
IVC	Inferior vena cava
L	Left
LA	Left atrium
LCA	Left carotid artery
LPA	Left pulmonary artery
LSA	Left subclavian artery
LV	Left ventricle
LVOT	Left outflow tract
MAPCAS	Major aortopulmonary collateral arteries
MV	Mitral valve
O	Oesophagus
PA	Pulmonary artery
PAIVS	Pulmonary atresia with intact ventricular septum
PLSVC	Persistent left superior vena cava
PV	Pulmonary vein
R	Right
RA	Right atrium
RCA	Right carotid artery

RVOT	Right ventricular outflow tract
RPA	Right pulmonary artery branch
RSA	Right subclavian artery
RV	Right ventricle
STIC	Spatio-temporal image correlation
SVC	Superior caval vein
T	Trachea
TAPVC	Total anomalous pulmonary venous connection
TGA	Transposition of the great arteries
TR	Tricuspid regurgitation
ToF	Tetralogy of Fallot
TV	Tricuspid valve
V	Ventricle
VSD	Ventricular septal defect

List of Videos

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Video 8.51 This video clip shows normal (blue) flow out of the left ventricle. There is retrograde filling of the branch pulmonary arteries

Video 9.1 Four-chamber view demonstrating ventricular disproportion. The right atrium and ventricle are wider than the left atrium and ventricle. The left ventricle does reach the apex however

Video 9.2 Sweep from the three-vessel to three vessel and tracheal view showing arterial disproportion. The pulmonary artery is larger than the ascending aorta and the arterial duct is larger than the transverse aortic arch, which is difficult to appreciate on 2D

Video 9.3 Sweep from the left ventricular outflow tract view to the three vessel and tracheal view showing arterial disproportion. The pulmonary artery is larger than the aorta and the arterial duct is larger than the transverse aortic arch. Color flow mapping shows there is antegrade flow across the slender transverse aortic arch

Video 9.4 A modified view showing the short axis of the heart. In the center of the picture is a bicuspid aortic valve

Video 9.5 Sweep from the four-chamber view to the three vessel and tracheal view showing ventricular and arterial disproportion. A dilated coronary sinus can be seen on the four-chamber view. The three-vessel view shows 4 vessels—the fourth vessel is the persistent left superior vena cava. The transverse aortic arch is slender

Video 9.6a Three vessel and tracheal view demonstrating a right sided aortic arch. The arterial duct passes to the left of the trachea and then behind it to join with the aortic isthmus on the right

Video 9.6b Three vessel and tracheal view with color flow mapping. This demonstrates antegrade flow in the right aortic arch and the left arterial duct

Video 9.7 Three vessel and tracheal view with color flow mapping demonstrating antegrade flow in both the dominant right aortic arch and the slender left sided aortic arch. The left sided arterial duct is also visualized and shows antegrade flow

Video 9.8 Sweep from the four-chamber view to the three vessel and tracheal view with color flow mapping. There is a normal four-chamber view with no regurgitation, the aortic arch is left sided with normal flow pattern and the origin of the right subclavian artery is normal, passing anterior to the trachea

Video 9.9a Modified three vessel and tracheal view using color flow mapping demonstrating a left sided aortic arch with an aberrant right subclavian artery passing behind the trachea. In order to obtain the aortic arch and the aberrant right subclavian artery in one view, the probe must be tilted

Video 9.9b Modified three vessel and tracheal view using color flow mapping demonstrating a left sided aortic arch with an aberrant right subclavian artery passing behind the trachea. In order to obtain the aortic arch and the aberrant right subclavian artery in one view, the probe must be tilted

Video 10.1 Transverse sweep from the fetal abdomen to the four-chamber view demonstrating the normal azygous vein. The small azygous vein is seen posterior and to the right of the descending aorta

Video 10.2 Parasagittal view demonstrating the course of the umbilical vein towards the ductus venosus on 2D and color. The usual course of the inferior vena cava toward the right atrium is also shown. At the origin of the DV, the blood coming from the umbilical vein accelerates leading to aliasing of the color flow signal

Video 10.3 Transverse view at the level of four-chamber view demonstrating the dilated coronary sinus and pulmonary venous flow both on greyscale and color flow Doppler. In cases where there is normal pulmonary venous drainage to the left atrium as demonstrated in this case, a dilated coronary sinus is suggestive of the presence of PLSVC

Video 10.4 Transverse view at the level of three vessel view demonstrating the presence of four vessels. From right to left, the right superior vena cava, ascending aorta, main pulmonary artery/duct and left superior vena cava are seen. The full length of the main pulmonary artery and duct is seen with the other vessels in cross-section

Video 10.5 Transverse view at the level of the three-vessel view demonstrating on color the presence of single left superior vena cava to the left of the pulmonary artery. On sweeping cranially, right to left flow in the innominate vein drain to the left SVC can be visualised

Video 10.6 Sagittal view of the fetal thorax and upper abdomen demonstrating the two parallel vessels of similar size “double vessel” appearance. The descending aorta is visualized in blue color and the azygous vein in red. No inferior vena cava is seen passing through the liver. Interruption of the inferior vena cava with azygous continuation is strongly associated with left atrial isomerism

Video 10.7 Sagittal view demonstrating direct drainage of the umbilical vein to the right atrium using color flow Doppler. This fetus had agenesis of the ductus venosus. The normal inferior vena cava has a course from the abdomen to the right atrium. Parallel to the spine the normal descending aorta is visualized (blue color)

Video 10.8 A transverse view at the level of four-chamber view demonstrating the smooth wall of the left atrium. No pulmonary veins are seen draining to the left atrium. A confluence is identified

at the back of the left atrium suggestive of total anomalous pulmonary venous drainage

Video 10.9a Transverse sweep from the four-chamber view to the three vessel view demonstrating the presence of an ascending vein (red color) at a place where a PLSVC is expected to be found

Video 10.9b In a more superior view the ascending vein is seen draining to the innominate vein (red flow) which joins the RSVC

Video 10.10 Tranverse sweep from the four-chamber view to an inferior plane where the dilated coronary sinus is seen. The pulmonary veins can be seen in red joining the coronary sinus and draining to the right atrium

Video 10.11 Fetus with infracardiac total anomalous pulmonary venous drainage. Longitudinal view of the fetal thorax and abdomen in color demonstrating the descending aorta and a venous channel originating from the confluence of pulmonary veins and draining into the liver

Video 10.12 Transverse view at the level of the three-vessel view of a fetus with left atrial isomerism. The left sided panel demonstrates dilated azygous vein draining into the SVC on 2D. This is confirmed on color flow Doppler on the right sided panel image. The dilated azygous vein (red color) draining into the superior vena cava is visualized. The aorta is seen in cross section and the pulmonary artery in blue color

Video 10.13 Transverse plane at the level of the four-chamber view demonstrating the presence of two vessels of similar size "double vessel" in front of the spine (the azygous vein and the descending aorta) which suggests the presence of interrupted inferior vena cava and left atrial isomerism. The azygous vein is posterior and to the right. The color Doppler flow demonstrates normal pulmonary venous drainage to the left atrium

Video 11.1 Atrial extrasystoles (atrial ectopic beats)

Video 11.2 Identification of supraventricular tachycardia using M Mode. The sample line is placed to ensure that it passes through atrial and ventricular tissue

Video 11.3 Supraventricular tachycardia with hydrops. Bilateral pleural effusions are seen. The atrial and ventricular rates are identical on the M-mode

Video 11.4 Atrial flutter. The atrial rate is much faster than the ventricles

Video 11.5 Fetus with ventricular tachycardia. The ventricular rate is faster than the atrial rate

Video 11.6 Demonstration of M mode to assess the atrial and ventricular rate simultaneously in the context of complete heart block. The M Mode cursor is aligned almost perpendicular to the atrial wall and the ventricle. The lower portion of the M-mode trace shows the regular atrial trace and the upper portion the slower ventricular rate

Video 11.7 Complete heart block due to maternal anti-Ro antibodies. There is good ventricular function and no effusions are evident

Video 11.8 Complete heart block due to maternal anti-Ro antibodies. This is seen in association with a pericardial effusion and widespread echogenicity of the heart at the crux, mitral valve and ventricular septum

Video 12.1 Axial bSSFP cine image series, equivalent to the "four-chamber view" in fetal echocardiography. The original dynamic sequence was acquired over several cardiac cycles, following which retrospective image processing techniques were applied to synchronise images to the correct cardiac phase and correct for gross fetal motion

Video 15.1 Four-chamber view of a fetus with non-compaction of the myocardium. The apices of both left and right ventricles are abnormally trabeculated with deep recesses (crypts)

Video 15.2 Four-chamber view of a fetus with non-compaction of the myocardium with the addition of color flow Doppler to demonstrate the deep crypts within the myocardium of the left and right ventricles which are pathognomonic of this condition

Video 15.3 This video shows a large cystic teratoma in association with fetal hydrops

Video 15.4 This fetus has a large teratoma with typical non-uniform echogenicity. It may be impossible to visualize the point of attachment of the teratoma to the heart or outflow tracts. Most are attached to the aorta

Video 15.5 This video shows the echogenicity of both the descending aorta and of the right pulmonary artery

Video 16.1 Four-chamber view of the fetal heart at 19 weeks gestation, demonstrating hypoplastic left heart syndrome (HLHS). The right ventricle is well developed and the tricuspid valve is seen to open normally. Sweeping to the well-developed arterial duct and hypoplastic aortic arch

Video 16.2 Four-chamber view of the heart demonstrating ventricular disproportion secondary to coarctation of the aorta

Video 16.3 Transposition of the great arteries. The four-chamber view and great arteries are balanced. The pulmonary artery arises posteriorly from the left ventricle and the aorta anteriorly from the right ventricle

Video 16.4 Four-chamber view of pulmonary atresia intact ventricular septum. The right ventricle is poorly functioning with evidence of endocardial fibroelastosis (EFE)

Video 16.5 Tetralogy of Fallot. The aorta overrides the ventricular septum and the pulmonary artery and arterial duct are significantly smaller than the left sided aortic arch

Video 16.6 Four-chamber view of the fetal heart. Complete atrioventricular septal defect (AVSD) with a balanced ventricular view

Video 18.1 Axial view at the level of the four-chamber view which demonstrates a thick atrial septum. On color there is only a small communication seen at the atrial level. This appearance is suggestive of restriction at the atrial level

Video 18.2 Balloon atrial septostomy. The balloon catheter is advanced from the IVC to the RA and then to the LA, where the balloon is inflated. Once clear of surrounding structures it is withdrawn sharply into the RA, tearing the atrial septum to create a generous interatrial communication. *IVC* inferion caval vein, *RA* right atrium, *LA* left atrium

Video 18.3 Transverse sweep from the four-chamber view to the three-vessel view in a fetus with Ebstein's anomaly. The typical apical displacement of the tricuspid valve is demonstrated and the atrialised portion of the right ventricle can be appreciated. There is no significant tricuspid valve regurgitation in this case. There is forward flow in the pulmonary artery seen. The size of the pulmonary artery is smaller compared to the aorta and this will be indicative of right ventricular outflow obstruction