

Feasibility, Technique and Potential Role of Fetal Cardiovascular MRI: Evaluation of Normal Anatomical Structures and Assessment of Congenital Heart Disease

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Abstract Fetal magnetic resonance imaging (MRI) is a third-level diagnostic tool for the study of fetal malformations and has been applied in the diagnosis and definition of fetal central nervous system (CNS) and other fetal, placental and uterine diseases. Recent developments of new realtime sequences during free breathing without cardiac triggering have established a potential role of MRI in the study of fetal heart: MRI can study the morphology using steady-state free precession (TrueFISP) sequences on sagittal, coronal and axial planes, orthogonally oriented to the fetal diaphragm and allows to identify the viscerotrial situs, the heart and its axis. It is also possible to perform a dynamic study, through the acquisition of cine-MR sequences with real-time steady-state free precession (SSFP) oriented according to the standard projections used in fetal echocardiographic scannings. At the moment, there is no evidence that short-term exposure to electromagnetic fields of 1.5 T or less harms the fetus. MRI can analyze the normal anatomy by transverse, long axis and angulated views to visualize the principal cardiac planes. There are recent evidences of a useful role of MRI in the definition of congenital heart disease (CHD). The study of fetal CHD can be made by direct signs such as volumetric abnormalities of the heart and of the cardiac chambers, abnormalities of the structure, thickness and signal intensity of the myocardial walls, anomalies of the cardiac axis orientation, defects of the ventricular and atrial septa and anomalies of the origin, size and course of the great arteries. The difficulty to recognize a "normal" anatomical structure in the reference projections, the increase of the vascular size before a vascular stenosis and the presence of cardiomegaly and pericardial effusion are instead considered as indirect signs of CHD are considered as suspect for fetal CHD. Despite current limitations, fetal MRI seems to be a promising diagnostic method for the assessment of the fetal heart.

Key Words: Fetal MRI, Fetal Heart, Congenital Heart Disease.

INTRODUCTION

Fetal echocardiography is at present the most commonly used diagnostic technique to detect congenital heart diseases (CHD). The application of echocardiography has completely modified diagnosis, counselling, management and least but not last prognosis of CHD, thanks to early characterization of these diseases in the prenatal period [1-3].

The main limitations of fetal echocardiography are well known and classifiable as intrinsic, like the direct operator experience and ability-dependence and ultrasonography equipment-dependence, and extrinsic, related to mother or fetus, like maternal habitus, oligohydramnios, gestational age and fetal position [4,5].

Thanks to recent technologic development and to the availability of the ultrafast sequences, it is now possible to use Magnetic Resonance Imaging (MRI) as III level method in the definition of fetal malformative diseases, after obstetrical screening and specialized ultrasonography performed in II level centres.

MRI examination allows a global evaluation of a case, through the analysis of both anatomy of the district of interest and its possible disruption expressed by modifications in signal intensity of parenchyma and affected organs.

In particular MRI may be useful in case of complex fetal anomalies or uncertain pathological conditions, ultrasonography (US) may not always allow to obtain adequate diagnostic information for therapeutic management of pregnant patient [6,7].

Thanks to high spatial resolution and the multiplanar imaging, MRI turns out to be suitable for documenting fetal diseases, particularly useful to study central nervous district and fetal body, as it has been recently reported in many

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papers of medical literature [8].

For a very long time, heart has represented an exception to the diagnostic possibilities of MRI above described, behaving like a real “black hole” [9]. This technical lack of fetal MRI (FMR) in representing fetal cardiovascular structures is due to three typologies of factors, concerning MRI method:

Small size of fetal heart and big vessels, which make the use of high spatial resolution sequences necessary; High heart rate, which requires high temporal resolution sequences; High blood flow, which requires high-quality sequences, not susceptible to moving fluids. Sequences usually used in FMR, single shot T2-weighted, cannot sample the signal coming from moving blood through heart and vessels and represent these structures as black, due to the absence of signal caught at that level.

On the other hand in adult patients MRI has become the gold standard for the characterization of anatomy, functionality and cardiac volumetry. On this basis some authors has recently begun to evaluate the potential role of MRI in the study of fetal cardiac diseases thanks to the use of new real-time free-breathing sequences without cardiac triggering and with good temporal and blood- myocardial contrast resolution, able to offer a representation of the cardiac dynamics and morphology [10].

SAFETY

MRI does not use ionizing radiations and up to now no detrimental effects have been shown for the fetus with exposition fields of 1.5 Tesla (T) or less. According to the last guidelines (Safety Committee of the Society for Magnetic Resonance Imaging), fetal MRI is recommended to be performed after the II trimester of pregnancy when other methods based on non-ionizing radiations are inadequate or inconclusive for the diagnostic characterization of the disease or if MRI is believed to provide critical information about therapeutic management and planning of pregnancy[11].

The main factors that can determine problems to the fetus are to be linked with three components of MRI equipment: magnetic field, radiofrequency and gradients. Queasiness, metallic taste and dizziness are the only side effects described for static magnetic fields up to 1.5 T.

More important are, instead, problems as amniotic liquid and fetus warming, noise and peripheral nervous stimulation caused by radiofrequency and gradients.

A rise of 2 °C or more in temperature has theratogenic effects to fetal central nervous system. However in vivo and in vitro studies on pregnant animals and human fetal samples do not show a meaningful temperature rise either on the maternal surface nor at fetal level, with a maximum rise value of 0.5 °C in 15 minutes and 1.5 T magnet studies. [12].

Neither a meaningful noise increase or auditory damages to children exposed to MRI in utero has been demonstrated, while amniotic liquid in fetal auditory canal has been indicated as protective factor against auditory damages [13].

Finally, turning on and off gradients during the acquisition of sequences determines mainly maternal skin surface warming, while the temperature decreases gradually when the distance from body margin increases.

The only general recommendation among fetal MRI operators is to alternate high SAR sequences with low SAR sequences during the examination, in order to let disperse, during this gap, warm that may had been created by high SAR sequences[14].

RESONANCE IMAGING AND STUDY TECNIQUE

Before performing every MRI examination an interview with the pregnant women is necessary, to ensure there are not common contraindications to MRI (cardiac pacemaker, metallic clips) and to collect clinical-

anamnestic data and to visualize ultrasound sonography and echocardiographic documentation where the pathological suspect originated. It is also important that medical staff obtains an informed consent; during this explanation a first approach have to be assessed with the patient, in order to obtain her active collaboration during sequences involving breathing and to explain fetal MRI performing procedures (duration, safety, diagnostic value).

MRI study has to be performed at least with an 1 T magnet, but the examination will be more diagnostic using a 1.5 T magnet. Different typologies of coils can be used depending also on the gestational age and sac length: phased-array or cardio multicanal surface coils are surely higher-quality because they allow to get higher signal also for limited longitudinal length (50-60 cm); you can also use integrated spine coils to study the body, since they allow to get a larger view during later gestational ages[15].

Pregnant patient lays comfortably and usually positioned supine or, if this position is not comfortable (compression of inferior vena cava, hydramnios, multiple pregnancies especially during later gestational ages), on the left side and she has to relax for a moment in this position in order to reduce spontaneous fetal movements.

In some cases, in order to minimize claustrophobic feeling, the patient can be positioned in the gantry in feet-first position. No sedatives or contrast agents are used either for fetus or for the mother.

If possible, it is recommendable to perform the examination in the morning, after a 4 hours fasting at least, since it has been proved that hypoglycemia reduces fetal movements; or, if ultrasound examination can be performed before MRI to assert fetal sleep or wakefulness stage, you can wait the sleep stage, taking into account that a regular fetal sleep-wakefulness cycle implies an alternation of this stages every 30 minutes.

Study protocol includes the acquisition of many sequences, some of them are necessary, others can be added optionally according to clinical request.

Images of each series are used as scout for the following sequences to minimize orientation problems owing to fetal position changes [16,17].

The main basic sequences used in standard fetal MRI are:

- A localizer sequence;
- A Single Shot (SSh) Half Fourier or FSE acquired T2-weighted sequence with maternal coronal plane for identification of fetal head, rachis and stomach and localization of placenta (anterior/posterior);
- A thin-slab (3-4 mm) SSh T2-weighted multiplanar-oriented sequence on axial, sagittal and coronal planes, orthogonal to the organ/district concerned, for a detailed evaluation of fetal anatomy.

The excellent compromise among spatial and contrast resolution and signal to noise ratio (SNR) of these sequences, as well as their execution speed, enable an outstanding visualization of fetal anatomy during every stage of pregnancy and, in particular, they allow to detect static fluids and structures featured by high fluid composition, like hyperintense structures, allowing then the study of fetal brain, of cavities containing fluid (nasal and oral cavity, pharynx, trachea, stomach and proximal intestine, urinary system, gall bladder), of lungs, placenta and amniotic fluid (AF). gradient-echo(GRE), 2D or 3D, breath-holding T1-weighted sequences, with and without adipose tissue signal saturation.

These sequences allow to recognize some organs and tissues thanks to the presence of blood, adipose tissue, meconium or other structures with high signal intensity on T1, enabling, in particular, to detect: thyroid or goitre or thyroidal ectopy; liver; intestinal meconium-filled loops (distal bowel), allowing differential diagnosis between urethral dilatation and megacolon; ischemic-hemorrhagic methemoglobin-filled areas and some encephalic structures like cortex, cerebellum and basal ganglia after the 24th week; placental vascular

stasis, haematoma or thromboses. Nevertheless, because of a longer acquisition time these sequences are more susceptible of artefacts caused by fetal motions [10,18].

Fetal cardiovascular study implies the use of the abovementioned thin-slab SS_H T₂-weighted sequences, as preliminary and anatomical sequences on three spatial planes and the acquisition of gradient-echo (GRE) steady-state free precession (SSFP) sequences on three spatial planes, to evaluate cardiac district and big vessels, according to the main views used in fetal echocardiography.

In fact, these sequences show an intermediate T₁ and T₂ contrast by using ultrashort TR (< 3ms), that is not susceptible of motions and allow to detect moving fluids as high signal intensity structures.

Cine-MR steady-state free precession (SSFP) with k-space sampling, radial and cartesian (2DFT) sequences, oriented according to standard views is also used in fetal echocardiography.

These sequences allow to detect heart and big vessels, thus obtaining the visualization of cardiac axis, evaluation of regular visceral-cardiac situs, localization of cardiac chambers and simultaneous atrial and ventricular contraction.

Combining SSFP with real-time technique has allowed the visualization of cardiac movement without triggering or synchronization with fetal heart or mother's breath-holding.

Usually an MRI study lasts from a minimum of 15 to a maximum of 20- 45 minutes.

ANATOMY AND ACQUISITION TECHNIQUE

The study of fetal heart with MRI technique is now possible by acquiring morphologic and dynamic sequences with multiplanar scans, using, in particular, two different techniques of anatomical planes acquisition [4,5].

The first technique is based on the acquisition of orthogonal view on three corporal axes (axial, coronal and sagittal) and multiple axial, sagittal and oblique scans, reproducing the main echocardiographic views, in particular [19,20]:

- Transverse views (four chamber view, origin of aorta (five chambers), pulmonary outflow tract (three vessels), aortic arch);
- Sagittal views (short-axis views of the left ventricle, tricuspid-aorta, long-axis of the ductus arteriosus, long-axis of the aortic arch);
- Oblique view (simultaneous visualization of long-axis view of the left ventricle, aortic arch and aortic duct).

The second acquisition method implies the use of views orthogonal to three corporal axes (axial, coronal, sagittal) and orthogonal to cardiac axes (long and short) following the technique usually performed in adult cardiac MRI [21]: in particular

- Orthogonal to cardiac long-axis (long horizontal and vertical axis of the heart, four chamber view);
- Orthogonal to cardiac short-axis (medium and basal short cardiac axis).

Both these techniques allow to determine heart position, its size, cardiac axis orientation, evaluation of location, position and size of cardiac chambers, main inflow vessels (SVC, IVC and pulmonary veins) and outflow vessels (Aorta, Ao, and Pulmonary Artery, PA), the concordance between ventricles and outflow vessels, aortic arch course and calibre.

Anatomical study of the heart implies systematic evaluation of cardiac orientation and volumetry, morphology and volumetry of cardiac chambers and the structure of myocardial walls, as well as the study of integrity of interventricular (IVS) and interatrial septum (IAS) with foramen ovalis (FO).

Anatomical study of big vessels, instead, implies evaluation of origin, calibre and course of cardiac outflow vessels (aorta, Ao, pulmonary artery, PA) and arterial duct (AD) and evaluation of calibre and course of inflow vessels (superior vena cava, SVC, inferior vena cava, IVC, pulmonary veins, PV).

In this study, we will refer to MRI views based on main echocardiographic views and a short MRI iconographic gallery will be shown, with anatomical structures that have to be visualized and analyzed on different acquisition planes.

TRANSVERSE VIEWS

Four Chambers (Figure 1)

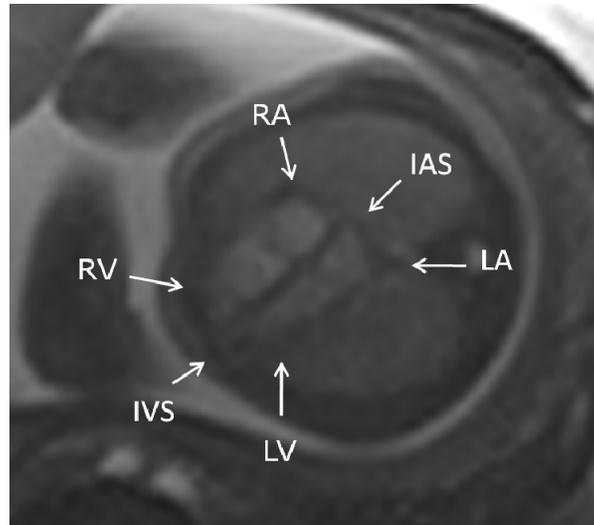


Figure 1: Showing the transverse view of the fetal thorax (RA right atrium, RV right ventricle, LA left atrium, LV left ventricle, IVS ventricular septum, IAS atrial septum)

Through this view we can analyse:

- Heart size compared to thorax (heart-thorax ratio)
- Position of cardiac apex
- Cardiac axis
- Structure of cardiac chambers
- Atrio-ventricular valvular plane (AVP)
- Ventricular and atrial septa

Four chamber plane evaluates first of all atrial and ventricular chambers sizes, apex orientation, cardiac axis angle (about 44°) and myocardial thickness. At the moment we cannot evaluate the difference of thickness relating to systole and diastole.

Myocardium shows a typical hypointense signal. In particular, myocardial wall of the left ventricle is thicker, more uniform and hypointense, while it appears thinner, jagged, porous in the right ventricle. It is not always possible to document the moderator band of right ventricle apex. There are many opposing opinions about it in literature. Its bad visualisation is an MRI limitation in assessing viscera-atrial situs. Ventricular septum is well visible and its thickness and signal intensity are equal to ventricular walls.

Ventricles appear basically symmetric, triangle-shaped, more strengthened the left one, more enlarged the right one. Papillary muscles can be detected especially when they are hypertrophic in a condition of myocardial hypertrophy.

Atrioventricular valves are only indirectly detectable and they appear like a thin hypointense line, called atrioventricular plane. Atrial myometrium is thin and symmetric and atrial septum appears as a hypointense line, better visible only after the II trimester of pregnancy.

Aortic Origin (Figure 2)

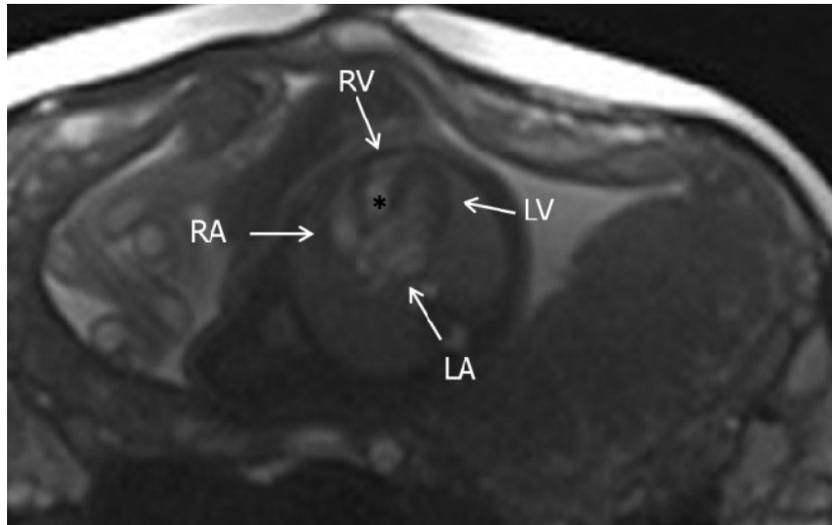


Figure 2: Five chambers; black asterisk Aorta origin

By this view, we can analyse the origin of the aorta from left ventricle, in the middle of the heart, as well as the structures of abovementioned views.

Pulmonary Outflow Tract (Figure 3)

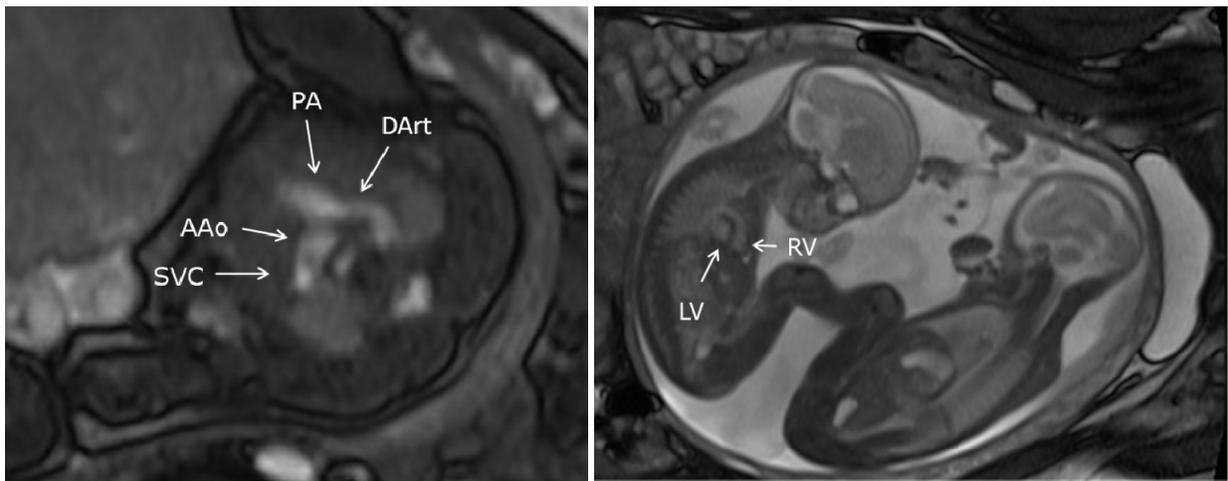


Figure 3: Three Vessels; SVC superior vena cava, AAo ascending Aorta, PA pulmonary artery-common tract, DArt Ductus Arteriosus)

It allows the evaluation, from right to left, of superior vena cava, aorta and pulmonary artery and the part of aortic duct that links PA to descendent Ao.

SAGITTAL VIEWS

Long-axis of the aortic arch (Figure 4)

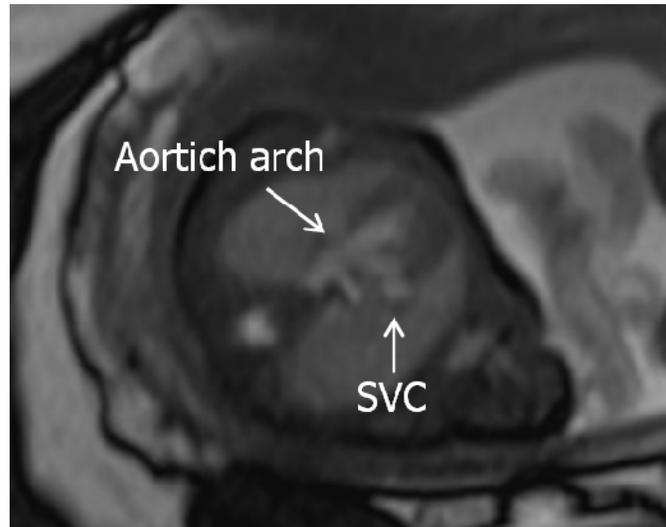


Figure 4: Long axis view of the aortic arch (superior vena cava SVC). This view shows both aorta arch and superior vena cava.

Short-axis view of the left ventricle (Figure 5)

Figure 5: Short axis view of the left ventricle

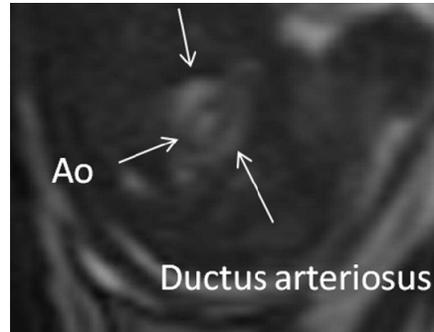
This view of the left ventricle allows a good evaluation of myocardial thickness, of the outflow tract of pulmonary artery and ventricles position.

Tricuspids-Aorta View (Figure 6)

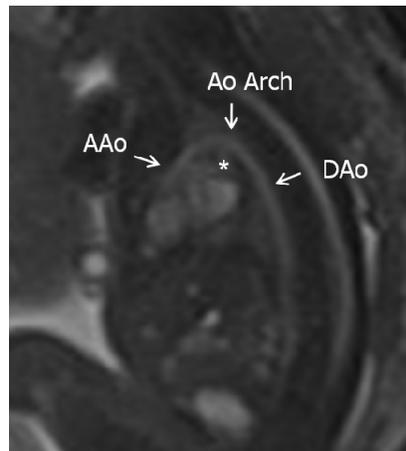


Figure 6: Tricuspids-aorta view (IVC inferior vena cava).

It allows to evaluate the position of right cardiac chambers and IVC and SVC inflow tract

Aorta Outflow Tract (Figure 7)**Figure 7:** Long-axis view of aortic duct

This view allows the visualization of PA that connects with Ao through aortic duct. This forms an arch that links the descending aorta.

Long-Axis View of the Aortic Arch (Figure 8)**Figure 8:** Long axis view of the aortic arch (white asterisk Pulmonary Artery)

This view shows the aortic arch on long axis with the origin of three epiaortic vessels.

OBLIQUE VIEWS**Long-Axis View of Left Ventricle (Figure 9)****Figure 9:** Long axis view of the left ventricle

This view visualizes outflow tract and the aorta at the origin and in its ascending tract.

Arch and Aortic Duct View (Figure 10)

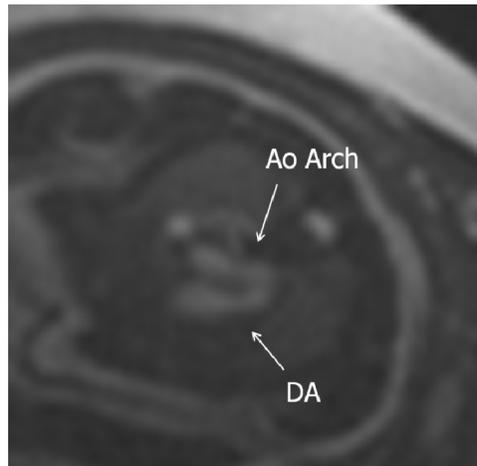


Figure 10: Arch and aortic duct view (DA ductus arteriosus)

It allows to visualize simultaneously arch and aortic duct and to compare their calibre and course.

Obviously, anatomical evaluation of heart and big vessels has to imply a simultaneous detecting of thoracic structures (thymus, lungs, trachea and oesophagus) and well detectable epiaortic vascular

STRUCTURES

Auxologic data about the evaluation of cardiac structures are not yet available, being the MRI study of fetal heart quite recent. Nevertheless, first results show that data are equivalent to echocardiographic biometry data. A summary of the main views is reported in Tab 1.

Table 1. Summarizing table of main MRI/echocardiographic views

MRI/ECHOCARDIOGRAPHY VIEWS		VISUALIZED ANATOMICAL STRUCTURES
Transverse	4 chambers	Heart size compared to thorax Position of cardiac apex Cardiac axis Cardiac chambers structure AV valvular plane Ventricular and atrial septum
	5 chambers	4 chambers + position and connection of the aorta at the origin with the heart Superior vena cava
	3 vessels	Aorta Pulmonary artery PA that connects to the Ao via the ductus, forming a ductal arch
	Long-axis of aortic arch	Ao arch and SVC myocardial thickness
	Short-axis of left ventricle	PA outflow tract Position of ventricles
Sagittal	Tricuspid-aorta	Position of right cardiac chambers IVC and SVC outflow tract Aorta outflow tract
	Long-axis of aortic duct	PA connects to descending Ao via aortic ductus
	Long-axis of aortic arch	Aortic arch on long axis with the origin of three epiaortic vessels Left atrium
Oblique	Long axis of left ventricle	Inflow tract and aorta at the origin and in its ascendant tract
	Arterial arch and aortic duct	Simultaneous view of Ao arch and AD arch

PATHOLOGY

According to the last studies published in literature, the fetal MRI of cardiovascular district and congenital cardiac diseases in utero is feasible and it is likely to be successful in the future, even though several technical limitations, only partially solved, still exist.

One of the first meaningful data about MRI study concerns its improved diagnostic accuracy during later ages of pregnancy compared to echocardiography, which is well known to be affected by limitations concerning the reduction of acoustic window (physiologic oligohydramnios) and gradual calcification in fetal ribs that impairs US propagation.

MRI study in cases of complex cardiac anomalies is certainly more difficult to arrange and understand: serious malrotation anomalies or severe ventricular hypertrophy may change normal heart geometry, thus requiring sequences oriented on new anatomical planes to be obtained during the examination itself. As a consequence longer acquisition time and sometimes sequences repetition until the correct standard anatomical view is obtained are necessary.

In MRI studies detailed knowledge of fetal cardiovascular district is fundamental and a training to learn how to perform and report an examination is necessary [22].

Some of the main limitations concern the impossibility to study by MRI cardiac rate alterations and to make hemodynamic evaluations of valve-based diseases (ex. incontinence, reflux), which can be only assumed once they have determined an anatomical alteration.

Cardiac contractility and valvular functionality, in fact, cannot be reliably evaluated by MRI. Technical impossibility of triggering fetal heart beat and still limited temporal resolution of current cine-MR sequences (2-3 frames per second) do not allow to analyse real-time cardiac functionality. The acquisition of cardiac movements is artificial, and their speed depends on the duration of acquisition of slices and not on the real velocity of atrioventricular movements.

In this research, we have tried to highlight the possibility to identify the main pathological aspects of congenital cardiovascular malformations and to create, starting from the experience and dictates of echocardiography, a new MRI semeiotics for the main congenital heart diseases.

MRI: Direct and Indirect Signs

The approach to the study of congenital heart diseases in echocardiography is usually based on sequential evaluation of anatomical structures and, in particular, it consists in building, during the examination, an anatomical sequence formed by atria, ventricles and great heart vessels, thus defining atrial situs, atrioventricular and ventricular-arterial concordances [23,24].

Classification of congenital heart diseases is based on anatomical-clinical aspects. According to these we can generically classify diseases in 4 main groups:

- With pulmonary hypoinflow (Pulmonary Stenosis, Tetralogy of Fallot, Tricuspid Atresia, Tricuspid Ebstein's Anomaly)
- With pulmonary hyperinflow (interatrial septal defects, IAD, interventricular septal defects, IVD, atrioventricular septal defects, truncus arteriosus, TA, and Aorto-Pulmonary septal defects)
- With pulmonary normal inflow (anomalous systemic and pulmonary venous returns, Ao coarctation, correct Transposition of the Great Arteries, aortic Stenosis)
- Duct-dependent (pulmonary atresia of ventricular septum (VS), Transposition of great arteries, mitral-aortic Atresia, Interruption of the aortic arch)

The correct approach to identify CHD requires first a screening examination, performed through routine obstetric ultrasonography, where 4 chamber view and outflow vessels view are visualized. In case of abnormal heart

visualization or in presence of specific risk factors for congenital heart diseases, depending on familiar, personal or fetal history, a II level examination such as echocardiography considered the gold standard for congenital heart disease diagnosis, is necessary. Echocardiography have to be performed in specialist centres by paediatric heart specialists or obstetric gynaecologists with specific training.

MRI methods can be defined as a III level method that only patients with echocardiographic suspect of cardiovascular disease can undergo [25].

MRI diagnosis of simple and complex malformative diseases of fetal heart is based on a integrated evaluation of direct and indirect signs of cardiac anatomy alteration.

We consider **direct** signs of cardiovascular disease the following aspects: morpho-volumetric anomalies of cardiac chambers and myocardium, malrotations, septal defects and anomalies of the origin, course and calibre of great vessels; while we consider **indirect** signs the absence of anatomical structures in the preliminary view, the prevascular calibre increase due to stenosis, the presence of cardiomegaly or pericardial effusion.

On the basis of these preliminary considerations we can classify MRI congenital heart diseases in three groups: diagnosable by the identification of direct signs, diagnosable by the identification of indirect signs or more often diagnosable by the identification of both typologies of signs [26].

Direct signs:

Ventricular septum defects both isolated and associated like in tetralogy of Fallot or truncus arteriosus are directly visualized as a solution of continuity of septum into infundibular, membranous or muscular part (Fig. 11), in particular by the acquisition of 4 chambers views and short-axis sagittal view of left ventricles. It is not always easy to distinguish an isolated IVD from other multiple and contiguous IVDs, especially in case of complex pathologic conditions.

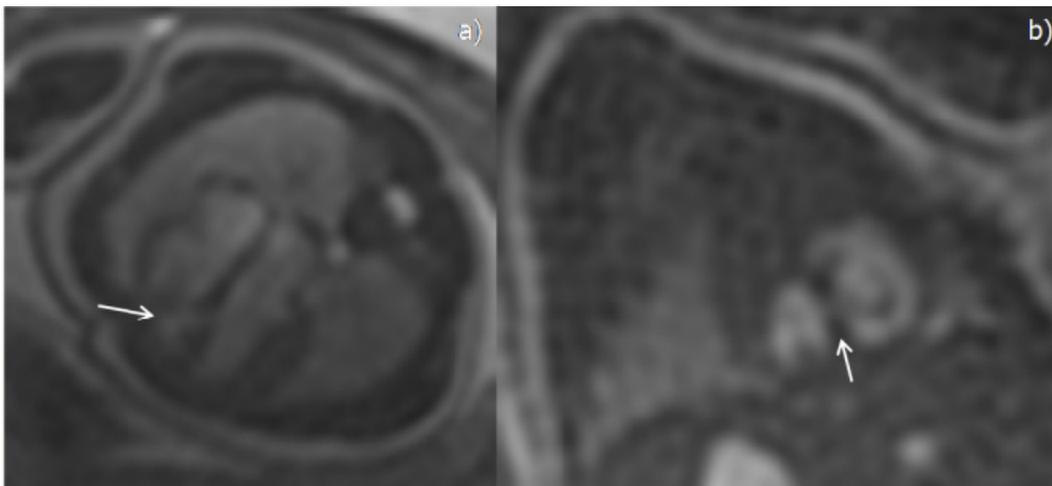


Figure 11: A millimetric solution of continuity of ventricular septum into muscular part, well showed by the acquisition of 4 chambers views (a) and short-axis sagittal view of left ventricle (b).

In complete atrioventricular canal, a great defect of IVS and IAS, for example, crux septum-primum, with disappearance or anomalous representation of atrioventricular plane, are considered direct diagnostic elements, even though valves cannot be visualized. Defining defect balancing or misbalancing depends on morpho-volumetric evaluation of two ventricular chambers and on the analysis of signal and myocardial thickness.

Hypoplastic left heart syndrome (Fig. 12) is detected also by MRI as a volumetric reduction of both cardiac left chambers till they become virtual volumes, with right ventricle forming the cardiac apex; there are also other direct signs concerning the calibre of Aorta, which appears particularly reduced at the arch level, and of the aortic duct, which appears increased in oblique view of the arch and aortic duct (in cases of duct-dependence of systemic circulation). Particularly useful are dynamic sequences that allow to detect small cavity of hypoplastic ventricle, that, due to its stuck walls, is sometimes undetectable with static sequences.

In situs inversus the position of heart and stomach on the right side is easily detectable, first of all by the position of fetus compared to the mother, thus defining right and left sides of fetus itself. In case of heterotaxic syndromes, instead, making differential diagnosis might be more complex.

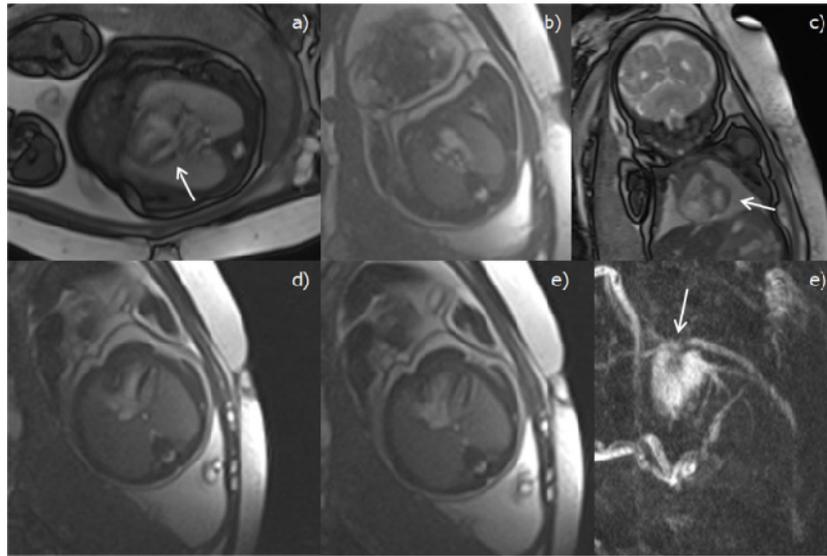


Figure 12: Hypoplastic left heart syndrome. Volumetric reduction of both cardiac left chambers till they become virtual volumes, with right ventricle forming the cardiac apex, on 4 chambers view (a) and coronal plane (c), in diastolic (d) and systolic phases (e); there are also other direct signs concerning the calibre of Aorta, which appears particularly reduced at the arch level (f) and of the aortic duct, which appears increased in oblique view of the arch and aortic duct (in cases of duct-dependence of systemic circulation) (b).

Anomalies of the origin (transposition of great vessels, overriding aorta, balanced truncus arteriosus) and course of great vessels (right-sided aortic arch) are all visualized by direct characterization of the vessel/s with anomalous origin or course. (Table 2, Figs. 13 and 14)

Table 2. Direct signs of morpho-volumetric anomalies

MORPHO-VOLUMETRIC ANOMALIES

Cardiomegaly

Left heart hypoplastic syndrome

VS defects (isolated and associated)

Tricameral biventricular heart

Situs ambiguous with right isomerism

Complete atrioventricular canal

Myocardial anomalies

(ex. Spungious cardiomyopathy, myocardial hypertrophies)

Left-malrotation

Right-malrotation

Tuberous sclerosis

(Multiple rhabdomyomas)

Pericardial effusion

DIRECT MRI SIGN

Cardiac- thoracic perimeters ratio inferior to 1/3

Volumetric reduction of both left cardiac chambers till they become virtual volumes and right ventricle forming cardiac apex

Solution of continuity of septum in infundibular, membranous and muscular part

One large atrial chamber and two ventricles

Stomach on the right side, transversalized medium liver, medium heart with apex on the left side and normal ventricles localization

Large VS and AS defect (crux septum primum) with disappearance of atrioventricular plane

Myocardial walls thickening compared to expected value considering gestational age according to normograms, acute signal hypointensity, more compact structure of myocardial wall, hypertrophic papillary muscles

Inclination angle of cardiac axis superior than 55° or inferior to 35° to meridian sagittal line

Multiple hypointense nodularities spread through myocardial walls

Hyperintense fluid pericardial > 2 mm thickness

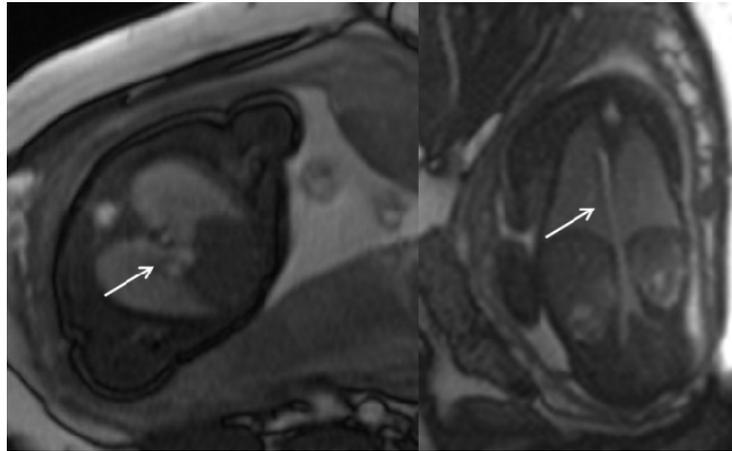


Figure 13: Right-sided aortic arch. The arch coursed from right to right directly posteriorly to join the descending Ao on the axial and coronal planes detected a right-sided aortic arch;

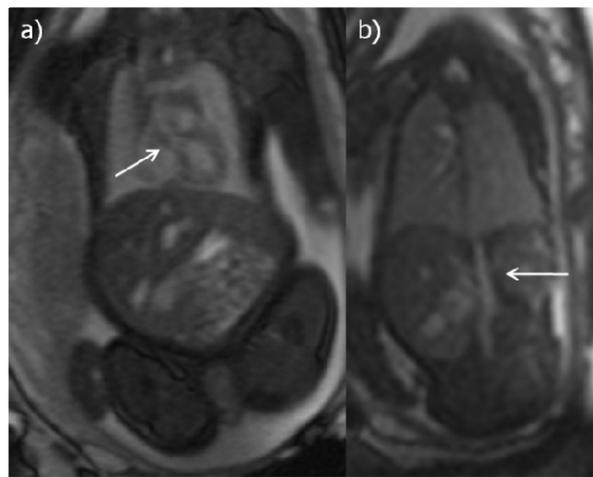


Figure 14: Right-sided aortic arch. Ascending Ao shows a normal origin and the descending tract crossed the midline at the level of diaphragmatic hiatus and lied just to the left of the abdominal spine.

In the transposition of the great arteries (TGA) (Fig. 15, 16 and 17) MRI allows to evaluate, by integrating dynamic and static sequences, the origin and the course of PA and Ao and ventricle-arterial discordance.

Defining a condition of correct transposition of great arteries (CTGA) might be more complex, especially during earlier gestational ages since it is more difficult to find the moderator band and consequently RV; through direct and indirect signs, instead, frequent associated anomalies, as, for example, large IVD and VA plane anomalies (direct) and right outflow tract obstruction (indirect), can be documented better.

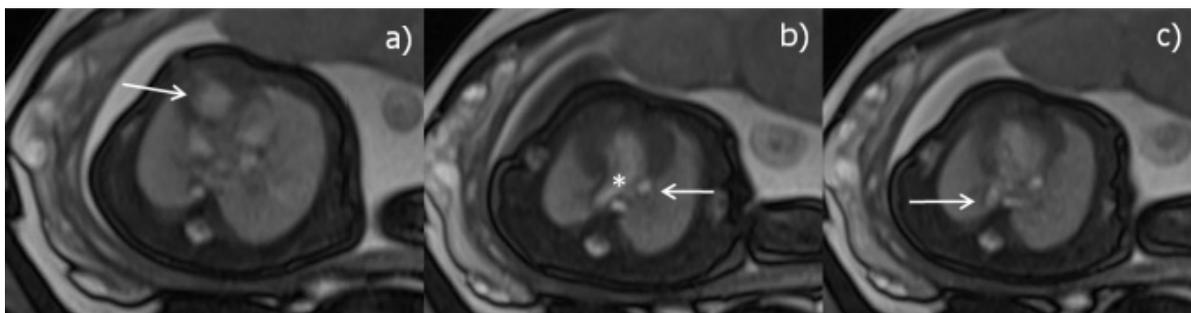


Figure 15: Complete transposition of the great vessels (TGA) with left ventricle hypoplasia and PA atresia. Transverse planes show the Ao (white arrow) arising from the RV (a), continuing into the arch (b, white asterisk Aortic arch, white arrow superior vena cava) and the descending tract that lies on the correct left side of the spine (white arrow in c), direct signs of TGA;

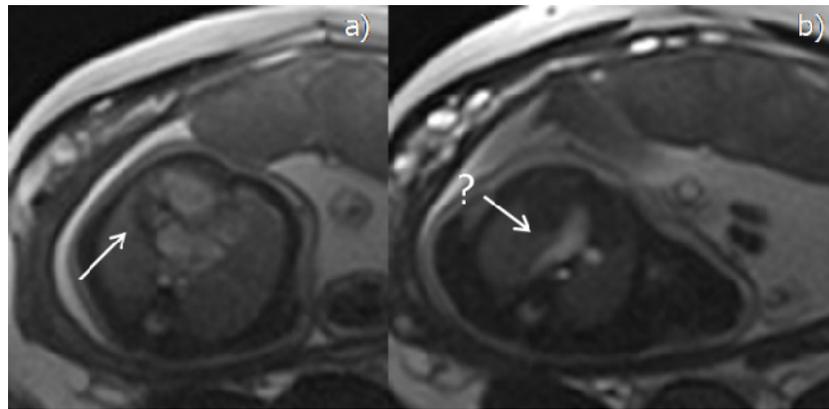


Figure 16: Complete transposition of the great vessels (TGA) with left ventricle hypoplasia and PA atresia.) Left ventricle appears particularly reduced of volume (white arrow in a) and on the three vessels view is not visualized PA and ductus arteriosus, indirect signs of PA atresia (white arrow in b)

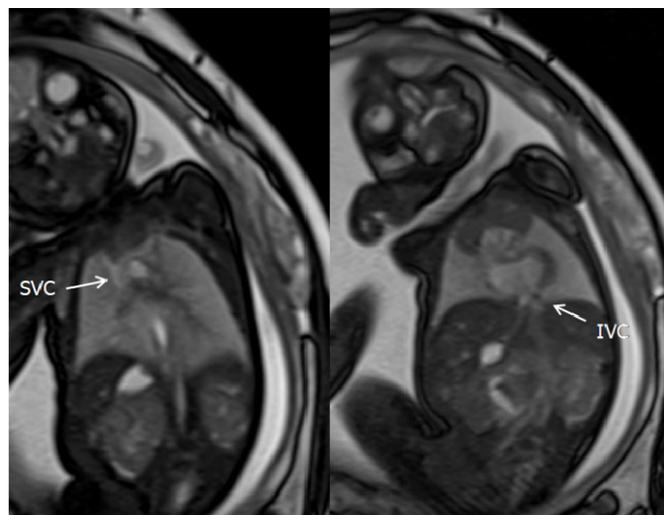


Figure 17: Complete transposition of the great vessels (TGA) with left ventricle hypoplasia and PA atresia.) Normal inflow in the right atrium of SVC and IVC with increased calibre

Also in Truncus arteriosus, through the integration of direct and indirect signs, we can define better direct visualization of a single cardiac outflow vessel (Fig 18 and 19) as well as the general pathological condition, by characterizing, in particular, the right position of the aortic arch and its potential interruption, its agenesis or AD dilatation, potential IVD (direct) and truncal valve stenosis that determines dilatation of ventricular chambers (indirect).

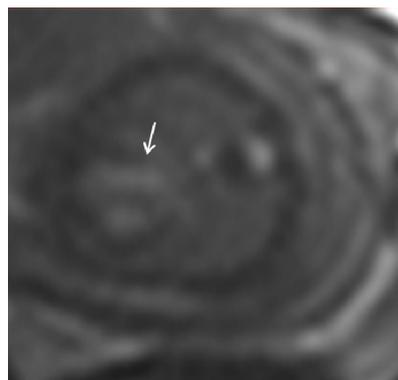


Figure. 18: Truncus Arteriosus type 2. The image on axial planes shows: a unique efflux vessel overriding the ventricles (truncus arteriosus, TA, white arrow).

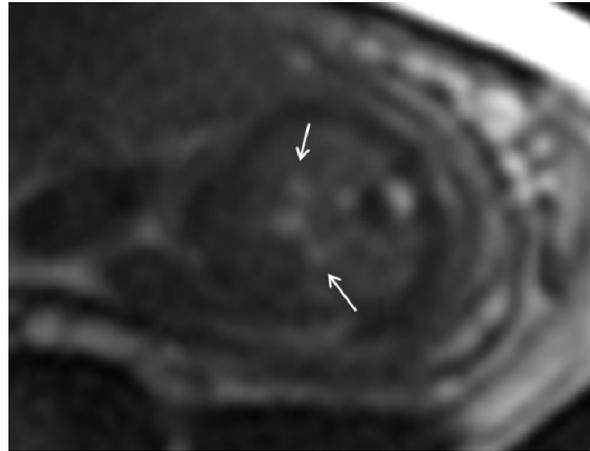


Figure. 19: Truncus Arteriosus type 2. The image on axial planes shows: a unique efflux providing both the lungs (right and left pulmonary artery) (white arrows) and continues the same way of the aorta.

The main direct signs of hypertrophic cardiomyopathy are: increase of myocardial walls thickness compared to expected value, considering gestational age (measured from endocardiac surface to epicardiac surface), acute signal hypointensity and more compact structure of myocardial wall, together with hypertrophic papillary muscle; the diagnosis of this primitive disorder of cardiac muscle is proved by echocardiography, which excludes mechanic and valvular causes.

The anomaly of origin and course of PA and Ao together with morpho-volumetric alteration of ventricles allows to recognize a condition of double-exit right ventricle (Fig. 20) and in particular: both outflow vessels origins mainly from right ventricles, showing an anomalous course, basically parallel, which is often associated with discontinuity of atrioventricular plane and IVD.

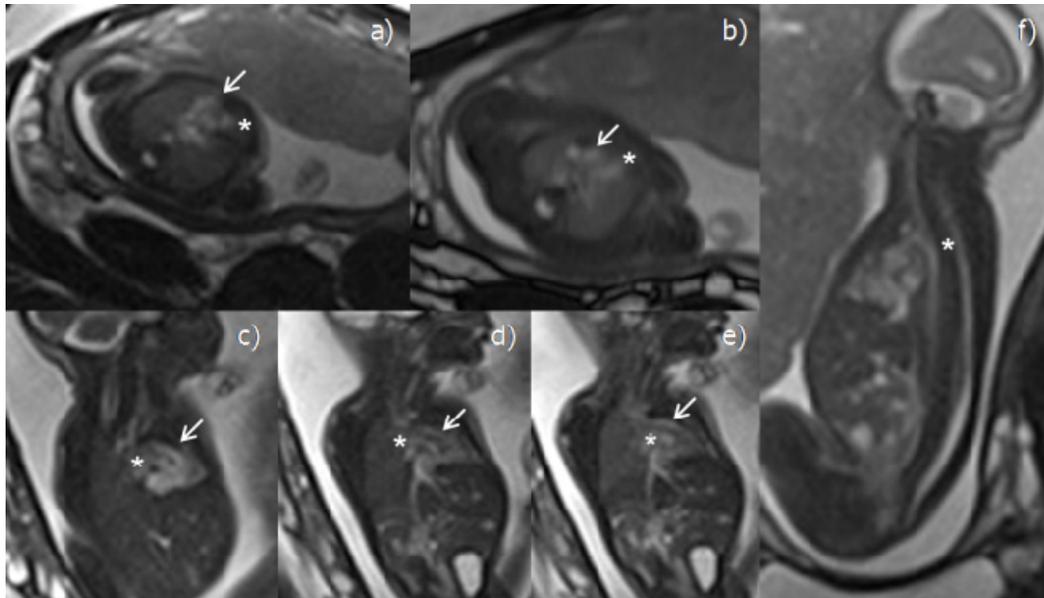


Figure. 20: Double-exit right ventricle. and in particular: both outflow vessels (white asterisk Aorta, white arrows PA) origins mainly from right ventricles, showing an anomalous course, basically parallel, which is often associated with discontinuity of atrioventricular plane and IVD.

In case of tuberous sclerosis, identification of common cardiac hamartomas (Fig. 21 and 22) by RMI is direct. They are spread more frequently in left ventricle or along IVS and with movable cardiac walls, as you can see in dynamic MR-cine sequences, and appear acutely hypointense in T2-weighted sequences and EGR sequences and hyperintense in T1-weighted sequences; moreover, thanks to multiplanarity and multiparametricity of the method, also subependymal nodules along cerebral ventricles and in Monro's foramen are easily detectable and identifiable.

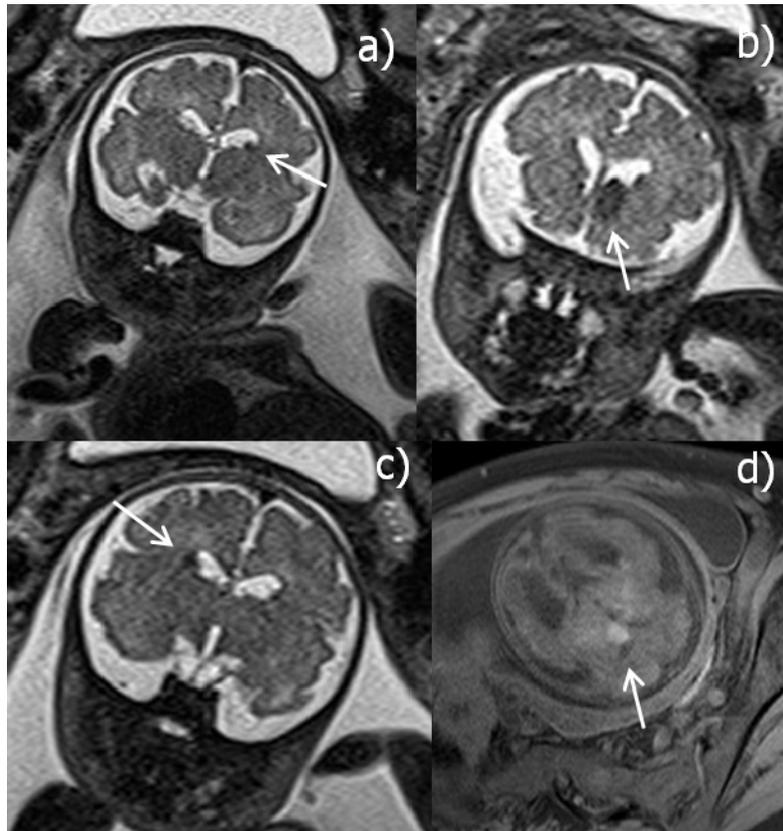


Figure. 21: Tuberosus sclerosis. The SSF T2 weighted images on the coronal plane (a,b,c) and T1 weighted with Fat Saturation image (d) on axial plane of the fetal head detect small subependymal nodules;

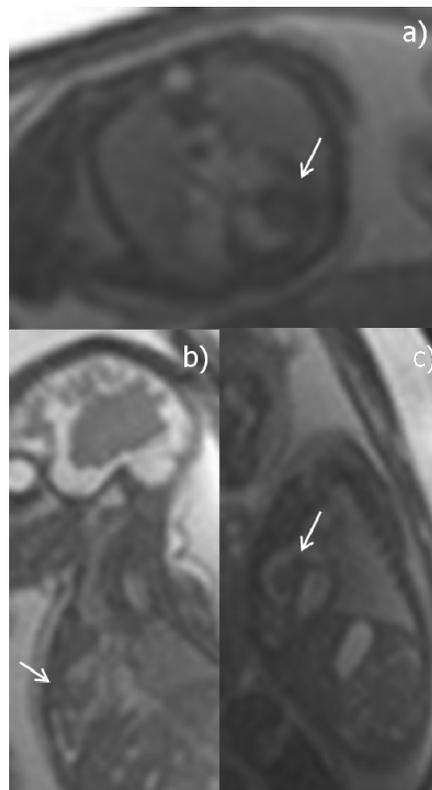


Figure 22: Tuberosus sclerosis The gradient-echo (GRE) steady-state free precession images on the transverse (a) and long-axis (b,c) planes show a big nodular hypointense lesion localized on the right Atrioventricular valvular plane (white arrows).

Indirect Signs

Vessels calibre anomalies and valves anomalies (Ao coarctation, pulmonary atresia/stenosis, mitral-aortal disease, tricuspid atresia) are basically revealed by indirect signs, in particular dilatation of cardiac chambers caused by vessel stenosis and post-stenotic dilatation. (Tables 3, 4)

Table 3. Indirect signs of valvular and great vessels anomalies

VESSELS CALIBRE ANOMALIES / VALVULAR ANOMALIES	INDIRECT SIGN
Pulmonary stenosis	RV dilatation and hypertrophy + lack of visualization of right outflow tract and PA
Tetralogy of Fallot Pulmonary Artery hypoplasia	RV dilatation + direct signs + reduction or lack of visualization of right outflow tract and PA
Coarctation of the Aorta	LV dilatation and bad visualization of ascendant Ao and Ao arch
Tricuspid atresia	RA dilatation
Mitral-aortal disease	LV volumetric reduction with myocardial hypertrophy and episodes of slowing and turbulence of blood flow

Table 4. Indirect signs of anomalies of the origin and course of great vessels

ANOMALIES OF THE ORIGIN AND COURSE OF GREAT VESSELS	MRI DIRECT SIGN
Balanced truncus arteriosus	One single arterial outflow vessel that overrides VS
Complete transposition of great arteries	Ao originates from RV and PA originates from LV in heart with normal atrio-ventricular concordance
Overriding aorta (Tetralogy of Fallot, IVD)	middle-placed origin of Ao, overriding VS and two ventricles
Right-positioned aortic arch	Aortic arch with right- to-left course

Aortic arch and PA hypoplasia are directly visualized by detecting with standard views corresponding vessels, characterized by small sizes compared to surrounding vessels, even though differential diagnosis excludes an outflow obstructive disease of corresponding ventricles.

Since we cannot rely on a correct MRI evaluation about blood flow direction and characteristics, characterizing correctly this homogeneous group of heart diseases is more difficult.

In aortic coarctation it is possible to determine different typologies of obstruction related to the origin of epiaortic vessels (distally to left subclavian artery, between common left carotid artery and subclavian artery and between left common carotid and right truncus arteriosus) especially associated to indirect calibre increase or decrease.

The main indirect signs of diagnosis of aortic stenosis are volumetric alteration of left ventricle, potential dilatation of right ventricle, myocardial wall thickening, generalised hydrops and growth retardation.

In pulmonary atresia/stenosis conditions the main MRI signs are volumetric alteration of right ventricle (mainly increase), together with right atrium widening and myocardial hypertrophy, reduction or lack of visualization in preliminary views of right outflow tract and PA. (Fig. 23, [video 1](#))

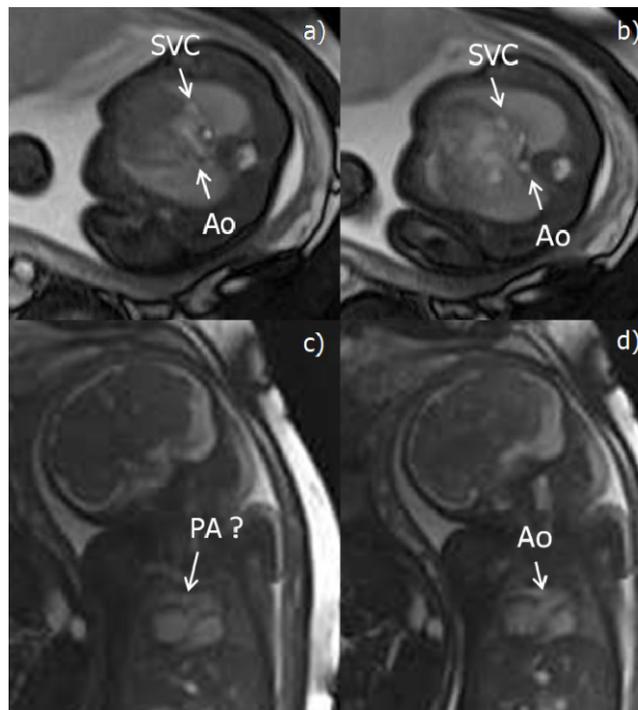


Figure. 23: Volumetric widening of right ventricle and atrium with lack of visualization on axial and coronal views of right outflow tract and PA (indirect signs)

It is important to notice that in every gestational age the evaluation of smaller vascular structures, like pulmonary veins, is more difficult, probably because of limited spatial resolution, and it is also more difficult to detect cardiac outflow vessels during earlier gestational weeks, probably because of their small sizes and because of increased fetal motion during MRI acquisition noticed during these gestational ages.

CONCLUSIONS

The use MRI in evaluating fetal heart still reveals some diagnostic limitations secondary to two typologies of factors: intrinsic and extrinsic.

The first intrinsic limitation concerns technical problems: it is still necessary to develop the equipment in order to overcome some lacks such as the low spatial and temporal resolution. Because of these limitations it is not possible to reproduce or give direct information about valvular or rate diseases.

The second extrinsic limitation is the still limited experience that is based on the few data still available in the literature. Multicentric studies are therefore necessary to acquire more experience, to construct biometric reference limits and generate diagnostic guidelines.

In the immediate future the use of dedicated sequences, with potential pseudo-angiographic study, might open new horizons. Similarly the application of new 3T magnet field equipments might increase spatial resolution. New possibilities of therapeutic treatments during prenatal period will undoubtedly require further efforts to reach higher image quality.

These will be our future challenges.

At the moment MRI can be performed in accurately selected cases, for unsure or multi-organ disease, where accurate counselling is necessary to plan clinic therapeutic iter of baby patient.

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