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IMAGES in PAEDIATRIC CARDIOLOGY

Hodzic A,[1] Maragnes P,[1] Dupont-Chauvet P,[2] Labombarda F.[1] Three dimensional echocardiographic assessment of multiple rhabdomyoma in newborn. Images Paediatr Cardiol 2013;15(4):1-4

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Abstract

Background: Cardiac rhabdomyomas represent the most common primary cardiac tumour in children and are strongly associated with tuberous sclerosis complex.

Results: We reported a newborn for whom three-dimensional echocardiogram, with multiplane mode, real-time imaging, full volume and i-slice view, allowed detailed visualisation of multiple highly echogenic and well-circumscribed cardiac rhabdomyoma.

Conclusions: Three-dimensional imaging allowed a better definition of the tumour characteristics and provided a better delineation of the spatial relationship of the mass with a tomographic perspective. Three dimensional imaging may facilitate a possible operative planning and should be included in cardiac mass evaluation and follow-up.

Patient

A male infant was born with a suspected diagnosis of tuberous sclerosis on antenatal ultrasound scan. At 5 days of age, three-dimensional echocardiogram (IE 33, 3DQ-QLAB advanced software, Philips®), with multiplane mode (Fig 1 - video clip 1 and 2), real-time imaging (Figure 2), full volume (Fig 3 - videoclips 3 and 4) and i-slice view (Figure 4 - videoclip 5) allowed detailed visualisation of three highly echogenic and well-circumscribed cardiac masses.

Figure 1: Three-dimensional echocardiogram (IE 33, 3DQ-QLAB advanced software, Philips®), multiplane mode.

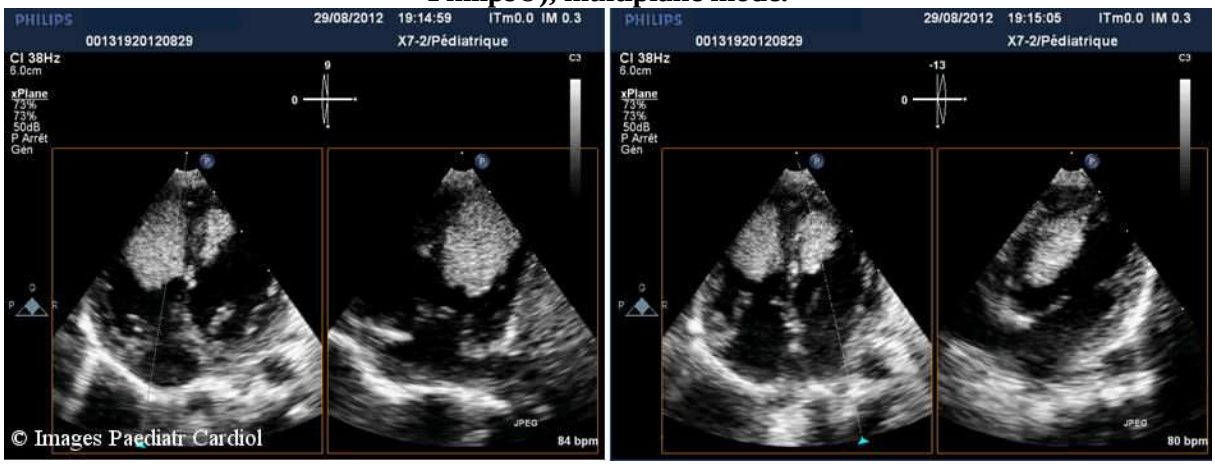


Figure 2: Three-dimensional echocardiogram (IE 33, 3DQ-QLAB advanced software, Philips®), real time imaging.

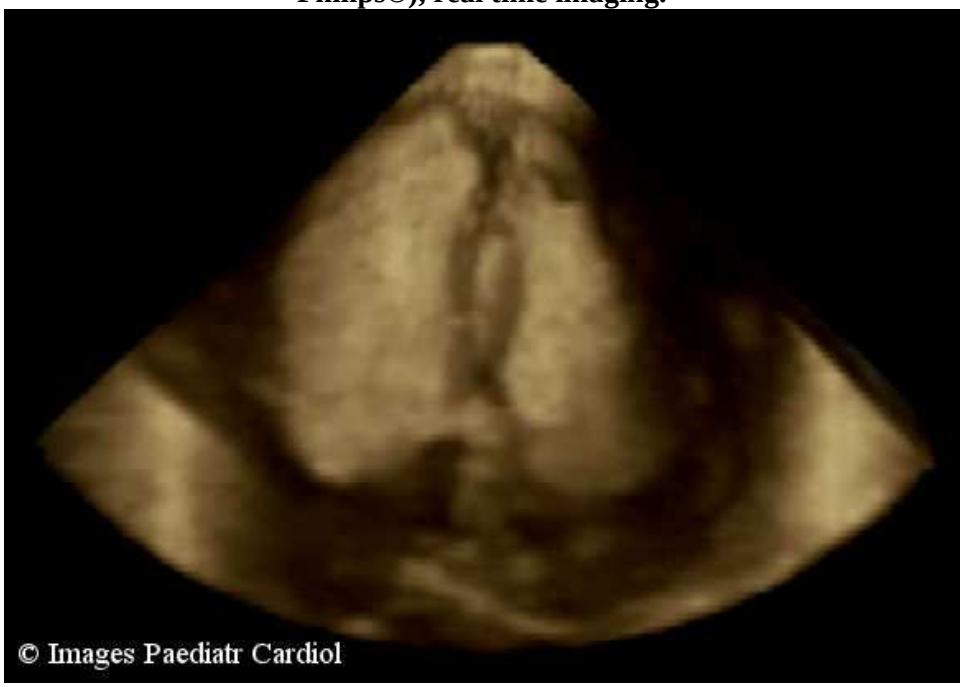


Figure 3: Three-dimensional echocardiogram (IE 33, 3DQ-QLAB advanced software, Philips®), Multiplane reconstruction.

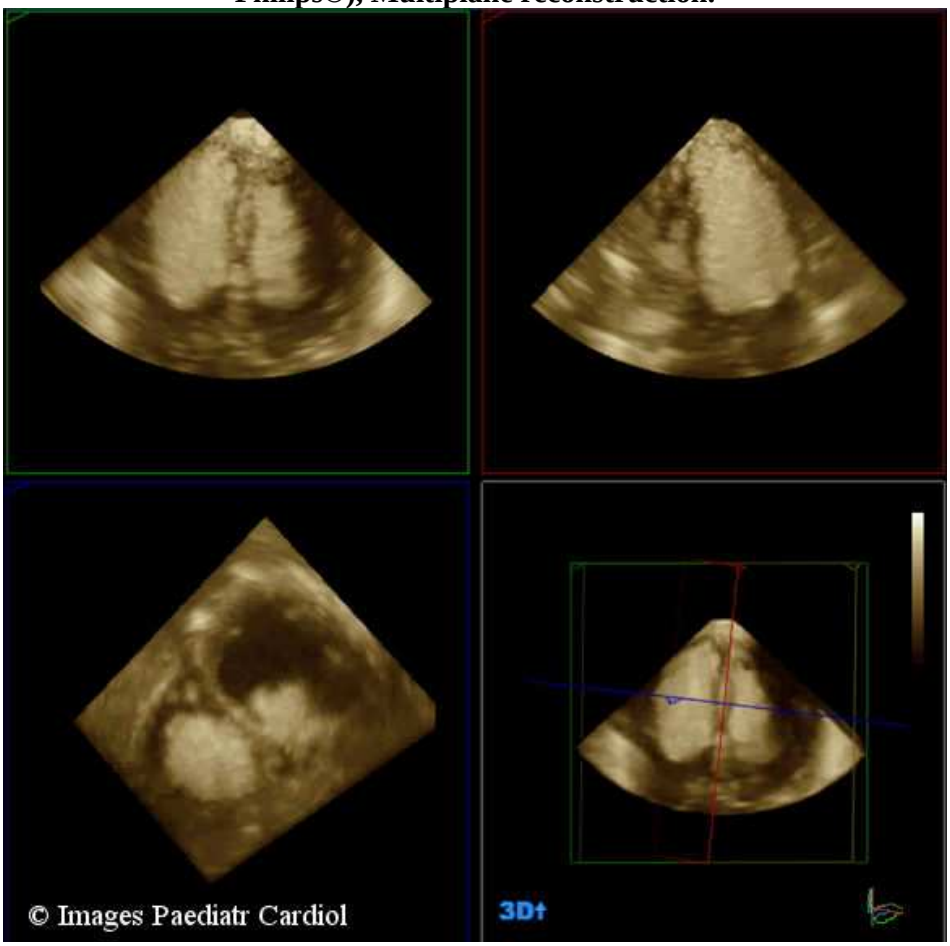
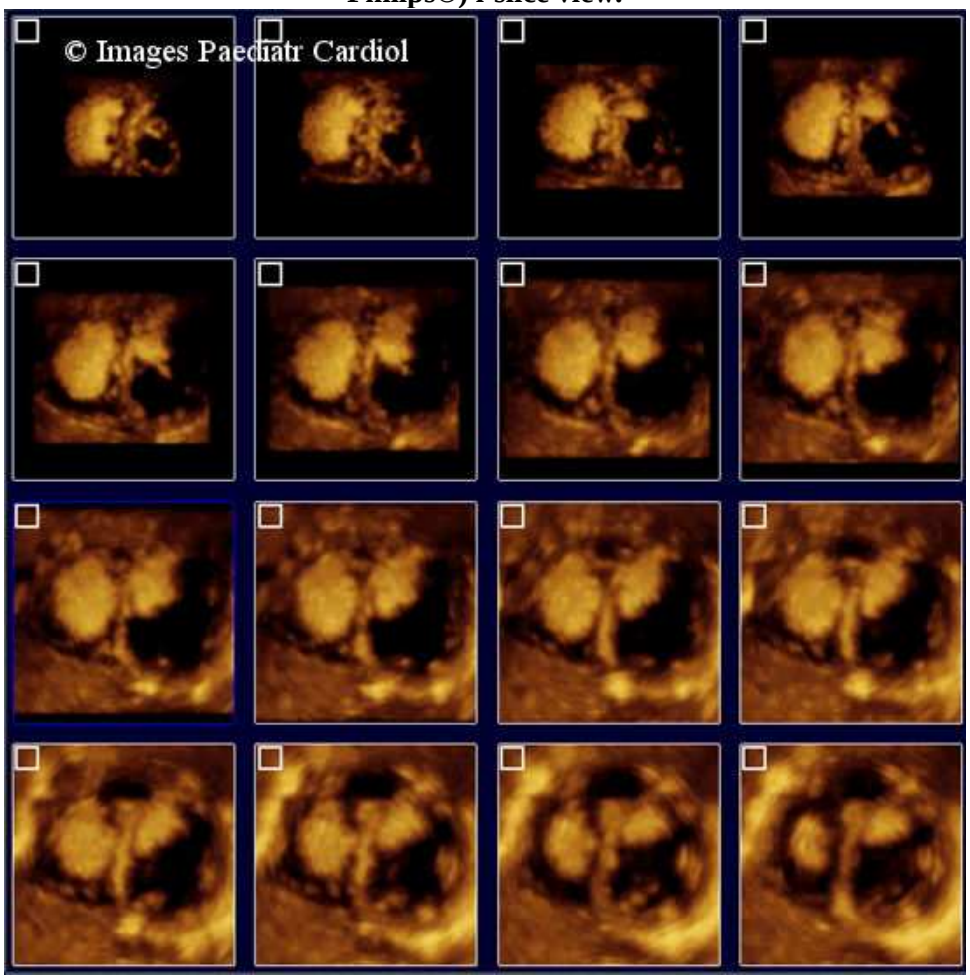


Figure 4: Three-dimensional echocardiogram (IE 33, 3DQ-QLAB advanced software, Philips®) i-slice view.



The first large lesion filled almost all the right ventricular cavity. The second large lesion was attached by a broad base and rose from the midportion of the interventricular septum to the left ventricular apex. A third small tumour was inserted under the pulmonary valve without outflow tract obstruction or pulmonary regurgitation. Postnatal magnetic resonance image revealed one brain lesion confirming the diagnosis of tuberous sclerosis. EKG registered recurrent supra ventricular arrhythmias successfully treated with amiodarone. A conservative approach was adopted.

Cardiac rhabdomyomas are benign foetal hamartomas with no malignant potential. They represent the most common primary cardiac tumour in children and are strongly associated with tuberous sclerosis complex,[1] especially when they are multiple. Tuberous sclerosis complex is a rare multisystem autosomal dominant genetic disease characterized by the widespread development of hamartomas in different tissues affecting mainly brain, skin, kidney and heart. Cardiac rhabdomyomas may occur in any location in the heart but are more common in the ventricles, often intracavitary and usually multiple. Multiple intracavitary tumours are considered as an important marker of tuberous sclerosis and the diagnosis may be made even in the absence of histological confirmation.[2] Cardiac rhabdomyomas are the earliest detectable tumour in the tuberous sclerosis complex during the prenatal period.[3] They may be isolated or associated with other specific lesions. The antenatal diagnosis by ultrasound scan has an important place for genetic counselling and patient management, due to the high frequency of associated tuberous sclerosis complex.

Postnatally, they lead to a wide clinical spectrum, ranging from fortuitous echocardiographic detection to congestive cardiac failure, arrhythmia, or sudden death, depending on the location, size and number of tumors.[4] However, most neonatal cases remain totally asymptomatic. Most of these tumours will disappear spontaneously. Complete regression is more common in the first four years of life. Conservative approach is the rule in the majority of case. Surgical intervention may be indicated for patients with hemodynamic compromise or intractable arrhythmias, with a low operative risk.[5] Three-dimensional imaging allowed a better definition of the tumour characteristics and provided a better delineation of the spatial relationship of the mass with a tomographic perspective in our patient. Three dimensional imaging may facilitate a possible operative planning and should be included in cardiac mass evaluation and follow-up.

References

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