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Double-Chambered Right Ventricle or Sub-Infundibular Stenosis Assessed by Cardiovascular Magnetic Resonance

Philip J. Kilner,^{1,*} Burkhard Sievers,¹ Gerd P. Meyer,¹ and S. Yen Ho²

¹Cardiovascular Magnetic Resonance Unit and ²Department of Paediatrics, Royal Brompton Hospital Site of Imperial College, Sydney Street, London SW3 6NP, UK

ABSTRACT

This paper presents magnetic resonance findings in four adults with doublechambered right ventricle or sub-infundibular stenosis, a condition that is treatable by surgery, but which can be difficult to assess echocardiographically. Four patients referred for cardiovascular magnetic resonance in the last 2 years were identified from CMR findings, although not necessarily from previous echocardiography, as having sub-infundibular stenosis. We used multislice spin echo imaging, cine imaging in oblique sagittal planes, and phase velocity mapping. We performed spin echo imaging of a post-mortem heart without known structural abnormality to illustrate right ventricular myocardial morphology. Results in patients showed evidence of sub-infundibular muscular obstruction separating the hypertrophied inlet and apical portions of the right ventricle from a thin-walled, unobstructed infundibular region in each case, with a systolic jet originating at least 15 mm beneath the unstenosed pulmonary valve. In addition to previously described structural components contributing to stenosis—enlargement and/or displacement of the septomarginal trabeculation, septoparietal trabeculations or the moderator band—CMR suggested additional components: a right ventricular papillary muscle in one, an anteriorly bulging aortic sinus in one, and hypertrophied muscular ridges of the parietal wall of the right ventricle. Even in this small group of patients, the causes of sub-infundibular stenosis appeared to be varied and multi-factorial. The abilities of magnetic resonance to give unrestricted, multi-planar views of right

*Corresponding author. Fax: +44-207-351-8816; E-mail: p.kilner@rbh.nthames.nhs.uk

ventricular anatomy, movement and flow make it well suited for diagnosis and characterization of sub-infundibular stenosis, especially in adults.

Key Words: Magnetic resonance; Congenital heart disease; Right ventricle; Stenosis

INTRODUCTION

Magnetic resonance gives unrestricted views of the right ventricle and pulmonary valve whereas ultrasound rarely gives adequate access to these areas, particularly in adults. Double-chambered right ventricle, in which right ventricular stenosis occurs below the level of the infundibulum, is less common than stenosis at infundibular or pulmonary valve level, but it is an important variant which should be recognized and distinguished. As the sub-infundibular stenosis of double-chambered right ventricle may progress and present during adulthood^[1] when echocardiographic access is rarely adequate, magnetic resonance imaging has an important role in assessment of such cases. Accurate pre-operative diagnosis can help to decide whether surgical relief of the stenosis will be feasible via a right atrial or right ventricular approach without incision or replacement of the pulmonary valve.^[1-3]

METHODS

Four patients referred to our CMR unit in the last 2 years were identified, from CMR findings, as having subinfundibular stenosis. We define sub-infundibular stenosis or double-chambered right ventricle as systolic jet formation caused by a narrowing in the right ventricle located below the infundibulum, with a nonobstructive and nonhypertrophied infundibular region between the stenosis and the valve. Provisional referral diagnoses based on previous echocardiography or catheterization are shown in Table 1.

A single post-mortem heart specimen without known morphological abnormality and fixed by immersion in formalin was also scanned by spin echo magnetic resonance imaging. Post-mortem images showing muscular ridges and bands of the right ventricle are included for illustration of structures which, in displaced or modified form, may contribute to sub-infundibular stenosis.

Magnetic Resonance Imaging

Imaging was performed using a 1.5 Tesla Picker Edge (Marconi, Cleveland, OH) system (patients 1 and 2) or a 1.5

Tesla Siemens Sonata (Siemans, Erlangen, Germany) system (patients 3 and 4, and the post-mortem specimen). Multislice spin echo images were acquired in transaxial and coronal planes. Cine images were acquired in at least one oblique sagittal plane aligned with the systolic jet in the right ventricle. This was located either by aligning with respect to transaxial images, or by cross-cutting with respect to initial cine acquisitions until the jet was in plane. For cine imaging, a gradient echo sequence, TE = 14 msec, was used in the Picker system, and a TrueFISP sequence, TE = 1.6 msec, in the Siemens system.^[4] In three of the patients, phase velocity mapping, echo time = 3.6 msec, was used to measure peak velocity of the core of the jet in the right ventricle.^[5]

RESULTS

Figure 1 shows transaxial spin echo images through the right ventricle at levels above, at and below the stenosis in patient 1. Figure 2 shows systolic and diastolic frames from oblique sagittal cine images aligned with the jet which was found to arise in the right ventricle at least 1.5 cm beneath the pulmonary valve in each case. In patients 1 and 2 (imaged by gradient echo, TE = 14 msec) the systolic jet shows as signal void, whereas in patients 3 and 4 (imaged by TrueFISP, TE = 1.6 msec) the bright core of the systolic jet is outlined by dark lines of signal void that represent parajet shear regions.^[6] These cine images showed jet location in relation to adjacent structures, including the ventricular septum with its ridge known as the septomarginal trabeculation, the left ventricular outflow tract or aortic root, and hypertrophied muscle of the parietal wall of the right ventricle.

In panel 1 (patient 1) of Fig. 2, at the tip of the black arrow, a muscle band appeared to pass through the sagittal plane and contribute to obstruction. In panel 2, a similarly located structure, this time lying obliquely in the sagittal image plane, appeared to contribute to obstruction. In panel 3, the systolic jet arose between hypertrophied muscle ridges of the septal and parietal walls of the right ventricle, whereas, in panel 4, the jet arose between an anteriorly bulging sinus of the aortic root and hypertrophied muscle of the parietal wall of the right ventricle.

| Outcome | Surgery to reduce muscular ridges and a patch to the anterior wall of the RV. The band was identified as a papillary muscle which had to be preserved. | No immediate intervention for right ventricular stenosis. | Awaiting surgery. | Awaiting surgery. |
|---|--|--|--|---|
| Structures Contributing to Sub-Infundibular Stenosis from CMR Appearances | Septal and parietal muscle ridges, beneath which a band appeared to pass left to right, horizontally. | Moderator band and inferior border of LVOT patch. | Septoparietal band and a discrete fibrous ridge. | Indenting aortic root, fibromuscular ridge and hypertrophied muscle of the parietal wall. |
| Peak Jet Velocity by Magnetic Resonance (m/sec) | 9 | 4 | Not measured | 5.8 |
| Referral Diagnoses | Previously suspected VSD. Right ventricular hypertrophy. Pulmonary or subpulmonary stenosis, 100mmHg gradient. | RVOT obstruction thought to be due to bulging LVOT patch. | Suspected acyanotic tetralogy of Fallot. | Infundibular or pulmonary valve stenosis. Perimembranous VSD. |
| Previous Investigations, and Surgery if Done | Echocardiography and catheterization | Five previous operations for aortic coarctation, LVOT obstruction and aortic valve stenosis. Recent echocardiography | Echocardiography | Exploratory operation aged 16 found no significant pulmonary valve stenosis. Recent echo- cardiography |
| Patient Number, Gender, and Age in Years | 1) F 46 | 2) M 23 | 3) M 64 | 4) F 55 |

Table 1 Patients Studied



Figure 1. Transaxial spin echo images in patient 1. RVOT = right ventricular outflow tract, which is surrounded by thin-walled infundibular muscle. The arrow points to a stenotic orifice bounded by hypertrophied myocardium. RV = right ventricle, hypertrophied beneath the level of stenosis.

Figure 3 shows spin echo images of the postmortem heart, with muscular ridges and bands labeled according to terminology used by Alva et al.^[7] The muscular structures may be relatively thick in this specimen due to shrinkage associated with postmortem fixing. In the patients, spin echo multislice images in transaxial and coronal planes, as well as the cine images and velocity maps, provided information on the level and nature of right ventricular obstruction. Slices from transaxial acquisitions distinguished the hypertrophied lower portion of the right ventricle, the level of mid-right-ventricular obstruction, and a nonhypertrophied, thin-walled infundibular region. Slices from coronal acquisitions, comparable to the image plane of Fig. 3, left, helped to identify a thickened septoparietal trabeculation, apparently contributing to sub-infundibular stenosis in at least two of the cases (patients 3 and 4), and possibly in all four. Structures identified by CMR as contributing to obstruction in each patient are listed in Table 1.

DISCUSSION

Magnetic resonance multislice imaging in transaxial and coronal planes, oblique sagittal cine imaging aligned with the right ventricular jet, and, in three of the four, phase velocity mapping, all contributed to diagnosis and evaluation of sub-infundibular stenosis in these patients. We did not acquire sagittal spin echo images, although in retrospect they could have helped with identification of muscular ridges of the septal and parietal walls of the right ventricle.

In none of the four patients had the nature of right ventricular obstruction been adequately determined before referral, in spite of echocardiography in all cases, a relevant catheter study in one, and earlier exploratory surgery in another. In patients 3 and 4, subsequent echocardiographic and catheter findings (in 4 only) were in accord with those of magnetic resonance.

The exact nature of right ventricular obstruction appeared to differ in each case, although recurring features were hypertrophy of septomarginal, septoparietal and parietal trabeculations, encroaching on the stenotic orifice from three sides. Obstructive contributions from septomarginal and septoparietal trabeculations and the moderator band accord with previous descriptions.^[1,7-11] The apparent contribution of a right ventricular papillary muscle in patient 1, and that of a bulging sinus of the aortic root in patient 4, are factors that may not have been described previously. In addition, comparison of the oblique sagittal cine images (Fig. 2) with the post-mortem oblique sagittal image (Fig. 3, right) suggests that hypertrophied trabeculations of the parietal wall of the right ventricle also contributed to subinfundibular obstruction. This is a factor which would be difficult to identify by ultrasound or catheter investi-



Figure 2. Systolic (left) and diastolic (right) cine frames in four patients with double-chambered right ventricle, numbered 1-4. Black arrows are located in the hypertrophied inferior portion of each right ventricle, pointing up to the origins of systolic jets in the thin-walled infundibular portions. White arrows point to the un-stenosed pulmonary valve, closed in diastole, in each case.



Figure 3. Muscle ridges, bands and trabeculations of the right ventricle on spin echo images of a post-mortem heart without known structural abnormality. The oblique coronal image (left) is located just in front of the ventricular septum, including signal from the ridge of the septomarginal trabeculation. The lower part of the moderator band slopes forward, out of this plane, towards the parietal wall. The oblique sagittal image (right) shows the transected ridge of the septomarginal trabeculations of the parietal wall of the right ventricle. Asterisks mark corresponding points on each image. LV = left ventricle, RV = right ventricle, RV = right atrium.

gation, and might also be hard to assess at post-mortem examination.

Our in vivo magnetic resonance findings suggest varied, multi-factorial causes of sub-infundibular stenosis, even in this small group of patients. It seems likely that one or more abnormal factors could initiate an obstructive process, which might progress through hypertrophy of additional muscle bands loaded by the initial obstruction. The abilities of magnetic resonance provide unrestricted, multi-planar views of right ventricular anatomy, movement and flow make it well suited for investigation of sub-infundibular stenosis in adults. Future studies in larger numbers of patients may throw further light on the variability and relative prevalence of structural abnormalities that contribute to sub-infundibular stenosis.

APPENDIX

Suggested protocol for assessment of double-chambered right ventricle

1. Preliminary scout images followed by multislice imaging in transaxial, coronal, and sagittal planes, covering the right ventricle and pulmonary valve. A breath-hold dark blood sequence such as HASTE or Tubo Spin Echo may be preferable to non-breath-hold spin echo, particularly in regions of suspected obstruction.

- 2. Cine imaging (FLASH or a steady state free precession sequence such as TrueFISP) should be acquired in an oblique sagittal plane aligned with the mid-right-ventricular orifice and the pulmonary valve. The aim is to show the origin of the jet in relation to structures the right ventricle and pulmonary valve. Sequences with longer echo times (>10 msec) show marked signal loss from turbulence. Short echo times (<6 msec) allow depiction of the jet core in relation to surrounding structures only if the slice is aligned exactly with the jet core.
- 3. If the previous cine was imperfectly aligned with the jet, an oblique coronal cine should be acquired, aligned with the suspected jet, and from this, a second oblique sagittal cine should be aligned with the core of the systolic jet.
- 4. Velocity mapping is performed through a plane transecting the core of the systolic jet, located immediately downstream of the orifice, velocity encoded in the slice-select gradient direction. Echo time (TE) should be 4 msec or less. The

"venc" (velocity encoding range) should be between 4 and 6 m/sec, depending on the severity of stenosis expected on the basis of previous cine image appearances, or previous Doppler echocardiography, if available.

5. Oblique left ventricular long axis cines in LVOT and four-chamber views, and a ventricular short axis stack may be acquired for assessment of biventricular function and to look for ventricular septal defects, which may co-exist with doublechambered right ventricle.

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