



BRIEF REPORT

Double Orifice Mitral Valve: A Report of Two Cases

Double orifice de la valve mitrale: Un rapport de deux cas

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ABSTRACT

BACKGROUND: Double orifice mitral valve is rarely reported; and no report of it was encountered in medical literature from Nigeria.

OBJECTIVE: To document the existence of double orifice mitral valve co-existing with situs in versus in our environment.

METHODS: Two young men who were initially diagnosed as cases of rheumatic mitral incompetence presented for echocardiography. The investigation was for the purpose of confirming the clinical diagnosis, and to determine the functional states. They both underwent 2D and M mode echocardiographic assessment.

RESULTS: At echocardiography, both cases turned out to have double orifice mitral valves. There were two orifices between the left atria and ventricles on gross appearance at 2 D echo; with multiple M shaped mitral valve tracings on M mode echo. The second case in addition, had situs in versus discovered on Chest X-ray and confirmed on abdominal ultrasonography.

CONCLUSION: Rare as it seems, double orifice mitral valve also occurs in our environment. It can also co-exist with situs in versus as a congenital abnormality. Since it is surgically remediable, all children with cardiac murmurs should be availed of echocardiography for diagnosis and early treatment to avert cardiac dysfunction. *WAJM 2007; 26(4): 323-325.*

Keywords: *Double Orifice Mitral Valve, Nigeria. Situs in versus, echocardiography.*

RESUME

CONTEXTE: Double orifice de la valve mitrale est rarement signalé, et aucun rapport de celui-ci a été rencontrée dans la littérature médicale à partir du Nigéria.

OBJECTIF: document de l'existence d'une double orifice valve mitrale co-existant avec situs en contre dans notre environnement.

MÉTHODES: Deux jeunes hommes qui ont d'abord été diagnostiqués comme des cas d'insuffisance mitrale rhumatismale présenté à l'échocardiographie. L'enquête avait pour but de confirmer le diagnostic clinique, et de déterminer les états fonctionnels. Ils ont tous deux subi en mode 2D et M échocardiographiques évaluation.

RÉSULTAT: À l'échocardiographie, les deux cas se sont dotés d'une double orifice valves mitrale. Il y avait deux orifices entre les oreillettes et les ventricules gauche sur l'aspect macroscopique à 2 D echo; M façonné avec de multiples tracés sur la valve mitrale mode M écho. Le deuxième cas, en plus, avait découvert versus situs dans le thorax de rayons X et confirmé sur l'échographie abdominale.

CONCLUSION: il semble que rares, double orifice valve mitrale se produit aussi dans notre environnement. Il peut également coexister avec situs en versus comme une anomalie congénitale. Comme il est réparable chirurgicalement, avec tous les enfants murmure cardiaque doit être fait usage de l'échocardiographie pour le diagnostic et le traitement précoce pour éviter un dysfonctionnement cardiaque. *WAJM 2007; 26(4): 323-325.*

Mots clés: *Double orifice de la valve mitrale, au Nigéria. Situs en versus, échocardiographie.*

INTRODUCTION

Abnormalities of the mitral valve be they congenital or acquired lead ultimately to congestive cardiac failure. Since they are surgically remediable, early identification is necessary if normal cardiac function is to be maintained. Most mitral valve diseases in our environment are acquired, and result from rheumatic process. Double orifice mitral valve is a congenital heart disease that is infrequently reported.¹ It occurs rarely in isolation; co-existing more often than not with atrio-ventricular canal defects.² Other malformations that have been reported include co-arctation of the aorta, bicuspid aortic valves and aneurysm of the right sinus of vasalva.³ No case has to my knowledge been reported from Nigeria, neither did one encounter a case co-existing with situs in versus in English medical literature. It is for these reasons that these cases are being reported.

CASE REPORTS

Case 1

Mr. A was sent for echocardiography from a peripheral centre. The indication was congestive cardiac failure secondary to rheumatic mitral incompetence. He was in his early twenties. On physical examination, he was orthopnoeic and oedematous with ascites, hepatomegaly and postero-basal crepitations. A grade 4/6 pan-systolic murmur was audible on auscultation of the praecordium over the mitral area. At echo, there were two valves between the left atrium and the left ventricle. Figure 1 is the M mode tracing of the heart at the level of the mitral valve leaflets.

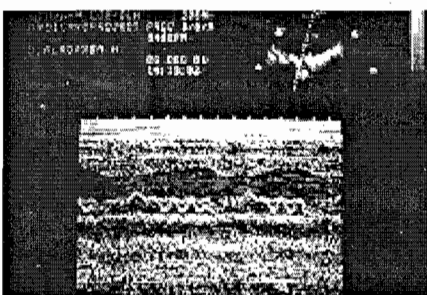


Figure 1: M mode echocardiography of the mitral valve. This picture shows the M mode image of the heart at the level of the mitral valve. There are multiple M shaped tracings instead of a single M shaped tracing usually recorded at the level of the mitral valve.

Case 2

Mr. S.A. presented in 2005 at the age of 14 years with a one-year history of exercise intolerance. He had no cough, orthopnoea, paroxysmal nocturnal dyspnoea, leg swelling, palpitation or chest pain. Up till the onset of his complaint, he had been active in school sports. There was no significant past medical history. On examination, he was lanky, but not Marfanoid. General examination revealed nil else significant. All systems were normal except for the cardiovascular system, where there was a bradycardia of 56/minute with ectopics. Blood pressure was normal at 120/70 mmHg supine. Jugular venous pressure was not raised. The apex beat was not displaced (5th left intercostal space, mid clavicular line). It was however heaving. Heart sounds were normal, but there was a grade 4/6 pan-systolic murmur loudest between the apex and the sternum. Rheumatic mitral incompetence was suspected. He was given Frusemide 40 mg daily and Slow K 600 mg daily; and then sent for investigations. On review after two weeks, he felt better and was tolerating more physical activity. Apart from mild hypokalemia, all blood investigations returned normal. Electrocardiogram showed sinus bradycardia with infero-lateral ischaemic changes, and voltage evidence of left ventricular hypertrophy. At echocardiography, there was left ventricular hypertrophy with the septum being slightly thicker than the posterior wall. There were 2 mitral valve orifices (Figures 2 and 3). Postero-anterior chest X-ray showed gastric bubble under the right diaphragm. The situs in versus was confirmed on abdominal ultrasound as the stomach was shown wedged between the liver and the right kidney. He was changed to Spironolactone 25 mg twice daily, and continues to do well even with follow-up.

DISCUSSION

Double orifice mitral valve in 50% of cases are detected during investigation of congenital heart disease.⁴ The remaining are discovered accidentally during surgical correction of congenital heart disease or autopsy.⁵ About 25% of the patients with double orifice mitral valve are said to present with mitral



Figure 2: 2-D long axis view of the heart. This is a 2-D echocardiographic image from the long axis parasternal view of the heart, showing two orifices connecting the left atrium and left ventricle.

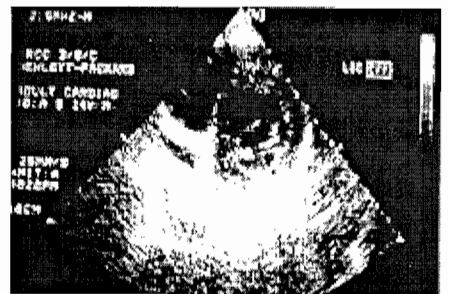


Figure 3: 2-D short axis parasternal view of the heart. This is a 2D echocardiographic image of the heart in short axis parasternal view; at the level of the mitral valve. The double fish mouth or a figure 8 tracing in the left ventricle depicts the two orifices.

incompetence as the dominant haemodynamic abnormality.⁴ Long standing mitral incompetence causes left ventricular dysfunction and ultimately right heart failure. Case 1 was already in congestive cardiac failure when he presented. He was in the third decade of his life. Case 2, being in his second decade was just beginning to decompensate. It should be expected that those cases with other structural heart defects would present earlier in life. The M mode picture showing multiple mitral valve tracing (Figure 1), the mitral valve orifices showing on long axis parasternal view (Figure 2) and short axis parasternal view (Figure 3) are confirmatory of double orifice mitral valve.

The lesion is amenable to surgery if discovered early. There is therefore the need to avail young children with cardiac murmurs of echocardiography in time, to avoid discovery after cardiac decompensation has set in. Even though

it is said that data on its incidence are lacking internationally,⁴ this treatise seeks to document the existence of this clinical entity among Nigerian Africans; and that situs in versus can be an accompanying congenital abnormality.

ACKNOWLEDGEMENT

I wish to thank the staff of our Medical Illustration and Technology Unit for assistance in producing the illustrative

figures. Drs. J. E. Ekedigwe and S. D. Pam, consultant Radiologists in our hospital are also appreciated for their opinion on the X-ray and ultrasound confirmation of situs in versus.

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