Interactive CardioVascular and Thoracic Surgery 25 (2017) 665–666
doi:10.1093/icvts/ivx164 Advance Access publication 5 June 2017

CASE REPORT

Cite this article as: Tagliasacchi I, Martinelli L, Bardaro L, Chierchia S. Minimally invasive mitral valve repair in osteogenesis imperfecta. Interact CardioVasc Thorac Surg 2017;25:665–6.

Minimally invasive mitral valve repair in osteogenesis imperfecta

Isabella Tagliasacchi, Luigi Martinelli, Leopoldo Bardaro and Sergio Chierchia*

Istituto Clinico Ligure di Alta Specialità (ICLAS), Rapallo, Italy

* Corresponding author. Istituto Clinico Ligure di Alta Specialità (ICLAS), 25 via Cabruna, 16135 Rapallo, Italy. e-mail: schierchia@gvmnet.it (S. Chierchia).

Received 19 December 2016; received in revised form 5 April 2017; accepted 22 April 2017

Abstract

Osteogenesis imperfecta is a disorder of the connective tissue that affects several structures including heart valves. However, cardiac surgery is associated with high mortality and morbidity rates. In a 48-year-old man with osteogenesis imperfecta and mitral valve prolapse, we performed the first successful mitral valve repair by right anterior mini-thoracotomy. At the 1-year follow-up, he was asymptomatic and echocardiography confirmed the initial success.

Keywords: Osteogenesis imperfecta • Mitral repair • Minithoracotomy

INTRODUCTION

Osteogenesis imperfecta (OI) is an autosomal heritable disorder caused by mutations of genes encoding for Type I collagen [1]. The syndrome presents with heterogeneous phenotypic manifestations that involve the skeletal system and several other structures including the heart [1, 2]. Valvular regurgitation along with aortic root dilatation are the most frequent abnormalities and mitral valve prolapse has been suggested to occur in up to 7% of patients [2]. Operative mortality and morbidity are considerably greater than in the control population and may reach 25% and 50%, respectively [2–4].

We here report the first successful minimally invasive valve repair in a man with OI and severe mitral incompetence.

CASE REPORT

A 48-year-old man with OI and severe mitral valve incompetence was referred to our outpatient clinic. His father was also affected by the disease and had died following an attempted mitral valve replacement complicated by acute cardiac rupture.

On physical examination, he had moderate cyphoskoliosis, blue sclerae and no signs of heart failure or pulmonary congestion. Echocardiography showed posterior mitral leaflet prolapse with severe regurgitation and normal ventricular function (Fig. 1). He had no coronary artery disease and was elected to undergo mitral valve repair.

To avoid sternal and parasternal lesions, a right anterior mini-thoracotomy was performed in the fourth intercostal space (Fig. 2). A number of precautions were used to avoid damaging the heart and surrounding tissues. (i) To prevent fractures, excessive rib spreading was avoided during exposure of the surgical field. (ii) Cardiopulmonary bypass was established by femoro-femoral

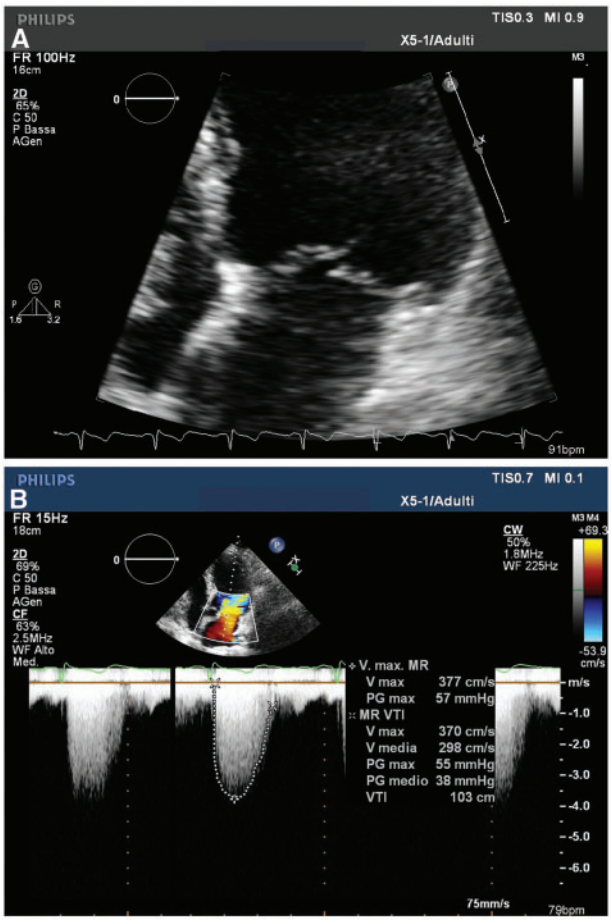

Figure 1: On apical 4 chamber view, the mitral valve appears thickened and mixomatous with prolapse of the posterior leaflet (**A**). Doppler imaging (**B**) shows severe mitral incompetence.

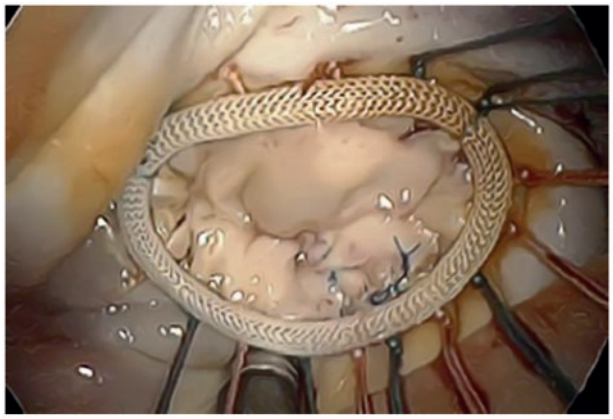

Figure 2: Intraoperative view. A #34 ring has been implanted on the annulus after P2 quadrangular resection.

artero-venous cannulation and cold cardioplegia was infused into the aortic root. (iii) As bleeding is virtually impossible to control before the unloading with the extracorporeal circulation, the right atrial wall was gently manipulated during this phase. (iv) Great care was taken in performing left atrial atriotomy, as the atrium appeared extremely frail. (v) Small and blunt needles were used to place stitches on the mitral annulus. (vi) Tissue resection was limited and folding was preferred when treating the prolapsing leaflets and (vii) two strips of pericardium were used to reinforce left atrial sutures.

The P2 prolapse was repaired by quadrangular resection and a #34 ring implant and intraoperative transoesophageal echocardiography showed no residual mitral incompetence. Complete haemostasis was obtained and no bleeding occurred postoperatively. Cross-clamp and cardiopulmonary bypass times were, respectively, 97 and 148 min.

The patient made an uneventful postoperative recovery and was transferred to the rehabilitation unit 4 days after surgery. At the 6-month follow-up visit, he was in good physical condition with excellent functional capacity and no mitral incompetence on repeat echocardiography. Similar findings were obtained at the 12-month control.

DISCUSSION

OI belongs to a group of heritable disorders of the connective tissue that includes Ehlers–Danlos, Marfan and Hurler syndromes. The phenotypic manifestations of the disorder are variable, affect several tissues, and may also involve the cardiovascular system [1]. Aortic insufficiency is the most common cardiac lesion, and mitral regurgitation the second [1–4]. Bonita *et al.* [2] reported 38 cases of OI with different types of cardiac involvement and 9 had mitral regurgitation.

In OI patients, cardiac surgery is associated with high mortality and morbidity rates that may reach 25% and 50%, respectively [4, 5]. In fact, as OI is a multisystem disease, several complications may occur in the perioperative period. These include respiratory infections, impairment of breathing mechanics, development of hyperthermia, prosthetic valve detachment and sternal and rib fractures. Furthermore, bleeding complications occur in 10–30% of cases [3–5]. Indeed, when surgical correction of severe aortic insufficiency was first attempted in 1965 by Criscitiello *et al.* [4], haemostasis was very difficult to achieve, and the patient died 4 days after the operation due to uncontrollable bleeding. Five (13%) of the 38 patients reported by Bonita [2] died from haemorrhagic complications and 6 of the 23 reviewed by Wong *et al.* [3], died because of severe postoperative bleeding.

For the reasons discussed so far, the surgical treatment of mitral incompetence should avoid sternotomy and valve replacement. Furthermore, as in our case, a number of specific manoeuvres should be applied for all surgical steps. Only 5 OI cases of mitral valve repair have been reported so far and all were performed by sternotomy [2, 3, 5]. To the best of our knowledge, this is the first patient where a minimally invasive thoracotomic approach was employed successfully.

Conflict of interest: none declared.

REFERENCES

[1] Radunovic Z, Wekre LL, Diep LM, Steine K. Cardiovascular abnormalities in adults with osteogenesis imperfecta. Am Heart J 2011;161:523.
[2] Bonita RE, Cohen IS, Berko BA. Valvular heart disease in osteogenesis imperfecta: presentation of a case and review of the literature. Echocardiography 2010;27:69–73.
[3] Wong RS, Follis FM, Shively BK, Wernly JA. Osteogenesis imperfecta and cardiovascular diseases. Ann Thorac Surg 1995;60:1439–43.
[4] Criscitiello MG, Ronan JA Jr, Besterman EM, Shoenwetter W. Cardiovascular abnormalities in osteogenesis imperfecta. Circulation 1965;31:255–62.
[5] Lamanna A, Fayers T, Clarke S, Parsonage W. Valvular and aortic disease in osteogenesis imperfecta. Heart Lung Circ 2013;22:801–10.