D. Ceroni
M. Beghetti
I. Spahr-Schopfer
A.A. Faundez
A. Kaelin

Corrective surgery for idiopathic scoliosis after heart transplantation

Received: 28 April 2000
Revised: 6 December 2000
Accepted: 18 December 2000
Published online: 3 March 2001
© Springer-Verlag 2001

D. Ceroni (✉) · A.A. Faundez · A. Kaelin
Paediatric Orthopaedic Unit,
Children's Hospital,
University Hospital Geneva,
Rue Willy Donzé 6, 1205 Geneva,
Switzerland
e-mail: dimitri.ceroni@hcuge.ch,
Tel.: +41-22-3824786,
Fax: +41-22-3824783

M. Beghetti
Paediatric Cardiology Unit,
Children's Hospital,
University Hospital Geneva,
Rue Willy Donzé 6, 1205 Geneva,
Switzerland

I. Spahr-Schopfer
Paediatric Anaesthetic Unit,
Children's Hospital,
University Hospital Geneva,
Rue Willy Donzé 6, 1205 Geneva,
Switzerland

Abstract Cardiac transplant surgery is being performed with increasing frequency as a treatment for end-stage heart disease. In addition to the well-known post-surgical problems of rejection and infection, these patients may present at a future date with other medical problems which require surgical treatment, including orthopaedic pathology. Severe idiopathic scoliosis has been described in association with congenital heart disease, and its surgical treatment poses considerable risks because of heart disease. Spinal fusion in heart transplant recipients involves similar risks due to the particular physiology and pharmacological reactions of the denervated heart. Several cases of cholecystectomy performed in heart transplant recipients have been described, but to our knowledge no orthopaedic procedures have been reported in such patients. We report on

a 15-year-old patient who underwent successful corrective surgery for idiopathic scoliosis 14 months after heart transplant.

Keywords Cardiopathy · Heart transplantation · Immunosuppressive agents · Idiopathic scoliosis · Spinal surgery · Risk assessment

Introduction

Cardiac transplantation in paediatric patients has been routinely performed in some institutions for more than a decade [10, 11, 15, 17, 20, 21]. The main indications are end-stage cardiomyopathy and congenital heart disease, with the latter being the most frequent. The population of paediatric heart transplant recipients is growing as their life expectancy increases due to improved immunosuppressive agents [10]. This new group of patients may also develop other diseases which might require surgical treatment. An association between congenital heart disease

and scoliosis is well established, and most of these cases involve idiopathic scoliosis without any morphological abnormality in the vertebral column [1, 3, 5, 8, 19, 22]. These curves usually develop quite early and are very severe, requiring surgical treatment [3]. With regards to patients with congenital heart disease, spinal fusion in post-transplant patients can be performed with a risk that varies according to the patient's preoperative cardiac status [3,8]. Indeed, post-transplant immunosuppressed patients are at a higher risk for septic complications that may compromise their life [13]. While there are published reports concerning urgent surgical treatment for acute cholecystitis or diverticulitis in such patients [2, 9, 12, 13, 18], there

is no mention of elective orthopaedic procedures in these individuals. We therefore wish to present our experience with corrective surgery for idiopathic scoliosis in a permanently immunosuppressed patient.

Case report

A 10-year-old girl presented with a juvenile idiopathic scoliosis characterized by a double thoracic curve (type V curve according to the King classification). At the age of 13 years and 10 months, she underwent orthotopic heart transplantation for end-stage cardiac failure (non-compaction left and right ventricle). While her cardiac function was normalized by the heart transplant, there was a significant worsening of the spinal deformity during her growth spurt, and corrective surgery for scoliosis was planned. Just prior to surgical correction at the age of 15, the spinal deformity revealed a 72° right thoracic curve and a 56° left cervico-thoracic curve. Bending radiographs showed a reductibility of the superior left thoracic curve of 28%, while that of the inferior right thoracic curve was 42% (Fig. 1). A multidisciplinary approach revealed that corrective surgery for her idiopathic scoliosis could be attempted. Echocardiography confirmed a good systolic and diastolic function without pulmonary hypertension, and endomyocardial biopsy revealed a low grade of rejection (ISHT II; International Society for Heart Transplantation rejection classification [4]). The spirometric and plethysmographic examination showed a uniform reduction of all lung volumes with a vital capacity of 1.1 l (40% of the predicted value). Erythropoietin administration with an iron-rich diet was ordered for the patient in order to increase blood mass for donation prior to surgery and to increase the hematocrit at the time of surgery. Immunosuppressive agents (0.17 mg tacrolimus/kg per day, 1 mg azathioprine/kg per day and 2.5 mg prednisone once a day) were continued to be administered even though we were aware that they might alter the patient's response to surgical stress and result in delayed healing.

The surgical procedure was performed under continuous transoesophageal echocardiography to monitor heart function. Continuous blood pressure monitoring by arterial line and central venous pressure monitoring were carried out. During surgery, the neurological status included somatosensory-evoked and neurogenic motor-evoked potentials. A posterior spinal fusion was performed from T3 to L3 using the Cotrell-Dubousset multi-segmented hook and pedicle screw instrumentation. As the laminae bone appeared very weak, no rotation or distraction manoeuvres were performed. After the spine had been exposed, the rods were moulded into the configuration of maximal reduction that could be obtained by manually straightening the spine. A cancellous bone graft was obtained by mixing bone from the laminae decortication with allograft (60 cc Tutoplast Spongiosa, Tutogen Medical, Neunkirchen). The surgical procedure lasted 4 h and 30 min, and blood loss was estimated at 1500 ml. Cell-saver permitted the recuperation of 300 cc of blood, and the patient received her predeposit autologous blood (2x500 ml) during surgery. Unfortunately, transfusion of 1000 ml irradiated and deleucocyted allogeneic blood was necessary. She received intravenous Tacrolimus and steroids for 48 h and was then placed on her usual p.o. regimen. Antibiotic prophylaxis (cefuroxime) was continued for 72 h and the patient was monitored in the intensive care unit for 3 days. She stood up for the first time on the sixth day without a brace and left the hospital on the tenth postoperative day.

Results

After surgery, the preoperative right thoracic curvature of 72° had improved to 33°, while the left cervicothoracic

Fig. 1 Pre-operative X-ray

curve had decreased from 56° to 36° (Fig. 2). Frontal and sagittal spinal balance were significantly improved by surgery. There were no problems with wound healing. Seventeen months after surgery, a solid fusion was noted without loss of the initial correction. Plethysmographic lung volume assessment showed a significant improvement of vital capacity, with an increase to 210 ml (+19.1%). In addition, the patient said she felt that she could “breathe better”.

Fig. 2 Post-operative X

Discussion

In spite of the improvement in surgical techniques and spinal instrumentation, corrective surgery for idiopathic scoliosis remains difficult and has the potential for serious complications. Patients with congenital heart disease undergoing general surgical procedures are recognized to have a significant rate of perioperative complications, and

we also suspect that spinal surgery in a heart transplant patient might be even more problematic [6, 7, 8, 16].

The denervated heart lacks the ability to respond acutely to hypovolaemia or hypotension with reflex tachycardia. Adaptation to stress is altered as the heart responds primarily by an increase in stroke volume. The increase in cardiac output therefore depends on venous return, with an initial increase in left ventricular end-diastolic volume, which in turn mediates an increase in stroke volume and ejection fraction by means of the Frank-Starling mechanism. In addition, there is a lag period in the haemodynamic response because the increased cardiac output is secondary to humoral reflexes (release of catecholamines) rather than to neural reflexes [14]. Heart transplant recipients are thus said to be “pre-load dependant” and require continuous echocardiographic monitoring of cardiac function perioperatively [6, 7, 8, 16]. In patients with congenital heart disease, the best predictor of perioperative complications is an American Society of Anesthesiologists (ASA) score of 3 or higher [8].

On the other hand, it is important to remember that the heart shows no response to drugs acting via the autonomic nervous system. However, it will respond to drugs acting directly on the heart or via humoral reflexes such as isoproterenol, ephedrine or dopamine [6, 7, 16].

The characteristics of the spinal procedure associated with perioperative complications include the length of the operation and the rate of blood loss, especially if a large amount of blood is lost within a short period of time [8]. There are many potential causes of excessive bleeding in this patient population. Immunosuppressive agents, such as corticosteroids, and inactivity are responsible for the osteoporosis that weakens the posterior vertebral arch, thus complicating fixation and increasing blood loss during surgery. In addition, immunosuppressive agents may result in delayed wound healing and possible wound dehiscence. An anterior approach is contraindicated to avoid re-entering the previously operated pleural cavity [8]. On the basis of these considerations, we believe that it is more reasonable to perform spinal fusion using a posterior approach only and to accept an incomplete correction with respect to the pre-operative bending radiographs. The rest of the treatment programme, including post-operative rehabilitation, should be exactly the same as for other idiopathic scoliosis patients.

Conclusion

The immunosuppressed state increases the heart transplant patient's susceptibility to infection, as it can alter the response to surgical stress and delay healing. Indeed, cardio-pulmonary function may be deteriorated at the time of surgery, thus increasing potential post-operative complications. However, orthopaedic elective procedures are not categorically contraindicated for these reasons, but do re-

quire certain precautions. The surgical technique has to be modified in order to decrease operative time and blood loss without compromising the surgical goals of correction and fusion. As the denervated heart is pre-load dependant, continuous transoesophageal monitoring is useful to pro-

vide information on heart filling. Our case report demonstrates that a necessary elective surgical intervention can be undertaken in a heart transplant recipient, but only after a multidisciplinary assessment of the patient.

References

1. Beals RK, Kenney KH, Lees MH (1972) Heart disease and idiopathic scoliosis. *Clin Orthop Rel Res* 89:112–116
2. Begos DG, Franco KL, Baldwin JC, Lee FA, Revkin JH, Modlin IM (1995) Optimal timing and indications for cholecystectomy in cardiac transplant patients. *World J Surg* 19(4):661–667
3. Beneux J, Rigault P, Pouliquen JC, Duval-Beaupere G, Pasteyer J, Durand Y (1976) Scoliosis and congenital cardiopathies. *Rev Chir Orthop Réparatrice Appar Mot* 62(8):781–792
4. Billingham ME, Cary NR, Hammond ME, Kemnitz J, Marboe C, McCallister HA, Snovar DC, Winters GL, Zerbe A (1990) A working formulation for the standardization of nomenclature in the diagnosis of heart and lung rejection: Heart Rejection Study Group. The International Society for Heart Transplantation. *J Heart Transplant* 9(6):587–593
5. Bitan F, Rigault P, Houfani B, Sidi D, Padovani JP, Merckx J, Durand Y (1991) Scoliosis and congenital heart diseases in children. A propos of 44 cases. *Rev Chir Orthop Réparatrice Appar Mot* 77(3):179–188
6. Boscoe M (1995) Anesthesia for patients with transplanted lungs and heart and lungs. *Int Anesthesiol Clin* 33(2):21–44
7. Cheng DCH, Ong DD (1993) Anaesthesia for non-cardiac surgery in heart-transplanted patients. *Can J Anaesth* 40(10):981–986
8. Coran DL, Rodgers WB, Keane JF, Hall JE, Emans JB (1999) Spinal fusion in patients with congenital heart disease. Predictors of outcome. *Clin Orthop* 364:99–107
9. Detry O, Defraigne JO, Chiche JD, Meurisse M, Joris J, Honore P, Jacquet N, Limet R (1996) Laparoscopic-assisted colectomy in heart transplant recipients. *Clin Transplant* 10(2):191–194
10. Fullerton DA, Campbell DN, Jones SD, Jagers J, Brown JM, Wollmering MM, Grover FL, Mashburn C, Luna M, Sondheimer HM (1995) Heart transplantation in children and young adults: early and intermediate-term results. *Ann Thorac Surg* 59(4):804–811
11. Hsu DT, Quaegebeur JM, Michler RE, Smith CR, Rose EA, Kichuk MR, Gersony WM, Douglas JF, Addonizio LJ (1995) Heart transplantation in children with congenital heart disease. *J Am Coll Cardiol* 26(3):743–749
12. Hull D, Bartus SA, Perdrizet G, Schweizer RT (1994) Management of cholelithiasis in heart and kidney transplant patients with review of laparoscopic cholecystectomy. *Conn Med* 58(11):643–647
13. Johnston TD, Katz SM (1994) Special considerations in the transplant patient requiring other surgery. *Surg Clin North Am* 74(5):1211–1221
14. Joshi GP, Tillmann Hein HA, Ramsay MAE, Foreman ML (1996) Hemodynamic response to anesthesia and pneumoperitoneum in orthotopic cardiac transplant recipients. *Anesthesiology* 85(4):929–932
15. Khaghani A, Santini F, Dyke CM, Onuzu O, Radley-Smith R, Yacoub MH (1997) Heterotopic cardiac transplantation in infants and children. *J Thorac Cardiovasc* 113(6):1042–1048
16. Lyons JB, Chambers FA, McSullivan R, Moriarty DC (1995) Anaesthesia for non-cardiac surgery in the post-cardiac transplant patient. *Ir J Med Sci* 164(2):132–135
17. McGiffin DC, Kirklin JK, Pearce FB (1997) Pediatric cardiac transplantation. *Adv Card Surg* 9:149–176
18. Menegaux F, Dorent R, Tabbi D, Pavie A, Chigot JP, Gandjbakch I (1998) Biliary surgery after heart transplantation. *Am J Surg* 175(4):320–321
19. Reckles LN, Peterson HA, Bianco AJ, Weidman WH (1975) The association of scoliosis and congenital heart defects. *J Bone Joint Surg [Am]* 57:449–455
20. Shaddy RE, Naftel DC, Kirklin JK, Boyle G, McGiffin DC, Towbin JA, Ring WS, Pearce B, Addonizio L, Morrow WR (1996) Outcome of cardiac transplantation in children. Survival in a contemporary multi-institutional experience. *Pediatric heart transplant study. Circulation* 94(9):69–73
21. Turrentine MW, Kesler KA, Caldwell R, Darragh R, Means L, Mahomed Y, Brown JW (1994) Cardiac transplantation in infants and children. *Ann Thorac Surg* 57(3):546–553
22. Voisin M, Dimeglio A, Pous JG, Dumas R, Jean R (1983) Spinal pathology and congenital cardiopathies. *Arch Mal Coeur* 76(5):568–574