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The classical and the one-and-a-half ventricular options for surgical repair in patients with discordant atrioventricular connections

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THE CLASSICAL OPTION FOR SURGICAL REPAIR IN patients with congenitally corrected transposition takes advantage of the physiologic correction provided by nature. At the end of the surgical procedures, however, the morphologically right ventricle remains as the systemic ventricle. Surgical intervention is essentially the correction of associated lesions, including closure of ventricular septal defects, pulmonary valvotomy, placement of a conduit from the morphologically left ventricle to the pulmonary arteries, replacement of the morphologically tricuspid valve, and placement of pacemakers for third degree atrioventricular block. For many years, the classical approach was the "standard" surgical approach.[1–4] More recently, newer alternatives have become available, including forms of anatomic repair, the "one-and-a half" ventricular option, and conversion to the Fontan circulation. The goal of anatomic repair is to craft connections such that the morphologically left ventricle becomes the systemic ventricle. Surgical techniques that accomplish this are a Rastelli procedure combined with an atrial baffle,[5] and the combination of an arterial switch with an atrial baffle, be it a Mustard or Senning procedure.[6]

The "one-and-a half" ventricular option is an intermediate technique between the classical repair and conversion to the Fontan circulation.[7] It is primarily indicated for patients with congenitally corrected transposition and pulmonary stenosis, with or without a ventricular septal defect. In this repair, the morphologically right ventricle remains the systemic ventricle, and pulmonary valvotomy is performed. Because in many of these patients pulmonary valvotomy alone would not be sufficient to decrease the left ventricular pressures to acceptable levels, a bidirectional Glenn is incorporated and this partially unloads the morphologically left ventricle. The goal, however, is to keep the left ventricular pressure sufficiently high to keep the septum in a normal location. This position of the septum leads to an improvement in morphologically right ventricular function, and decreases any tricuspid valvar insufficiency. For the properly selected patient, this also avoids placement of a conduit, which can be problematic in this population of patients. The alternative of conversion to the Fontan circulation is particularly appealing for patients with either complex intracardiac anatomy, which makes septation difficult, or pulmonary atresia, which requires placement of a conduit.[8]

Classical repair

There is indirect and direct evidence to support continued use of the classical repair. The indirect evidence is the long follow-up available from patients who have double discordance, but in the absence of associated lesions requiring surgical intervention. The direct evidence is the cumulative result of surgical series using the classical repair, for which there is also a long follow-up.

To cite some of the indirect evidence, Presbitero et al.[9] reported on 18 patients in whom the mean age at presentation was 35 years! Symptoms in 7 of these patients were caused by third degree atrioventricular block, in 9 by regurgitation across

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the morphologically tricuspid valve, and by supraventricular tachycardia, which occurred in all patients of age greater than 60 years. The patients were followed and the prevalence of complications in each decade of life was noted. Third degree atrioventricular block was the earliest complication and was observed in the first three decades of life in one-fifth of patients. Morphologically tricuspid regurgitation tended to begin occurring in the third to fourth decades of life. Arrhythmias and congestive heart failure, however, did not occur until the fifth and sixth decades of life. They concluded that patients with isolated congenitally corrected transposition have a good prognosis, especially for the first three to four decades of life.

Another study, from Toronto,[10] identified 52 patients greater than 18 years of age, and of these, 39 were surviving at an age of 32.7 years. Among those who died, mean age at death was 38.5 plus or minus 12 years, with half of the deaths occurring in patients less than 40 years of age. The most frequently noted causes of death were congestive heart failure due to morphologically right ventricular failure in 5 patients, and sudden death in 4 patients. This survival curve, although poor compared to a normal population, must be considered in the context of survival after paediatric cardiac surgery. Long-term survival has been analyzed in Finland following surgical correction of multiple different congenital cardiac procedures.[11] The survival curves for those with functionally univentricular hearts, and simple transposition, are not dissimilar from the survival curve noted for patients with congenitally corrected transposition in Toronto.

The direct evidence supporting ongoing use of the classical option for repair lies in the surgical reviews from several different centers that used this strategy in the past. These centres now have extensive long-term follow-up. Thus, 121 patients underwent surgery at Texas Children's Hospital between 1952 and 1999.[4] Of these patients, 47 underwent classical biventricular repair. The remaining patients had a potpourri of operations, including construction of a modified Blalock-Taussig shunt, banding of the pulmonary trunk, conversion to the Fontan circulation, placement of pacemakers, and so on. The operative mortality in the recent era was 2.5 percent. Survival at 10-years was 91 percent, and at 20-years was 75 percent. Risk factors for death included the presence of a common atrioventricular junction, and poor preoperative function of the morphologically right ventricle. At the Mayo Clinic,[3] surgical intervention took place in 111 children between 1971 and 1996 at a mean age of 9 years, with all patients undergoing a classical biventricular repair. A ventricular septal defect was closed in 101 patients, a conduit placed from the morphologically left ventricle to the pulmonary arteries in 65 patients, and 21 patients had replacement of the morphologically tricuspid valve. Early mortality in the last 10 years of the series, from 1986 to 1996, was 3 percent. At 10 years, two-thirds of the patients had survived.

A third large surgical experience was reviewed by the group from Toronto,[2] who performed surgery in 127 patients between 1959 and 1997. The majority had closure of a ventricular septal defect, in 95 patients, and treatment of pulmonary stenosis, with placement of conduits in 63 patients, and valvotomy or subpulmonary resection in 7. Third degree atrioventricular block was present in 12 percent of the patients, and 28 percent had significant atrioventricular valvar regurgitation, with the morphologically tricuspid valve replaced in 27 patients. The operative mortality for the first operative procedure was 6 percent. Late deaths, when occurring, were due to reoperation, sudden death, arrhythmia, and progressive heart failure. Of those patients only requiring closure of the ventricular septal defect, three quarters remained alive 35 years after the initial repair. The patients who had the poorest outcome were those who required replacement of the tricuspid valve. The investigators from Toronto also presented a meta-analysis of known reports of classical repair, accumulating 480 patients, with 13 percent operative mortality. Actuarial survival at 10 years for this cohort was 70 percent. The overall operative mortality following classical repair, therefore, can be quite low, varying from 2.5 to 4 percent. Late problems, nonetheless, such as morphologically right ventricular failure, atrioventricular valvar insufficiency, and third degree atrioventricular block, are interrelated and lead to a relatively poor survival in the long term. Thus, actuarial survival at 10 years is 74 percent, but at 20 years is no more than 48 percent. It is these results that must be compared to the results of other techniques such as the "double switch," the one-and-a-half ventricular option, and conversion to the Fontan circulation. A confounding issue, however, is that there is true long-term follow-up for the classical option, whereas follow-up for the other alternatives is relatively short.

Anatomic repair

It was these relatively poor outcomes over the long term that promoted the search for surgical alternatives. Anatomic repair has the goal of making the morphologically left ventricle the systemic ventricle. As already discussed, the two primary techniques are either the Rastelli procedure or an arterial switch combined with an atrial baffle.[5,6] The double switch is technically feasible in patients who have two functional arterial valves. The Rastelli option is appropriate for patients with a solitary functional arterial

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valve. The primary issue with both options is that they rely on operations already known to have significant complications over the long tern when applied to patients with simple transposition. The question is "do two wrongs make a right?"

First, with regard to the published outcomes of the anatomic repairs, in the Toronto experience with 127 patients with congenitally corrected transposition, 8 patients had the Rastelli operation, and one patient had a double switch.[2] One of the patients died, giving a survival of 89 percent. The authors noted that the double switch carried a higher operative risk than the classical repair. The combined potential complications of these procedures are significant. Obstructions and leaks are known potential complications of atrial baffling procedures, and atrial arrhythmias occur in essentially all patients if followed sufficiently long. Thus, in another series from Toronto,[12] the incidence of atrial arrhythmias at 25 years was greater than 60 percent[12] (Fig. 1). Late coronary arterial obstruction secondary to complications of transfer is a potential complication of the arterial switch operation, and obstruction of the right ventricular outflow tract can be the consequence of reconstruction of the pulmonary arteries. The Rastelli operation, furthermore, carries a risk of subaortic stenosis, which developed early in 3 patients reported from Toronto.[2] These patients may have also suffer ventricular dysfunction either from the baffle or from enlargement of the ventricular septal defect. Complete atrioventricular block can also be acquired in these patients. The meta-analysis reported from Toronto included data on 10 patients who had undergone a double switch, and 48 receiving the Rastelli option. Actuarial survival at 10 years was not available, but overall mortality at the most recent follow-up was 7 percent.

There are, nonetheless, several studies now of long-term follow-up of the Rastelli operation for simple transposition, all describing somewhat disappointing results. Boston Children's Hospital[13] reported survival of only half their patients after 20 years. In this series, sudden death, left ventricular dysfunction, myocarditis, and complications of conduits were reported as causes of death (Fig. 2). Of note, there was no difference in survival for the three different periods of 1973 to 1979, 1980 to 1989, and 1990 to 1998. Long term results are also reported from the Mayo Clinic.[14] Survival at 20 years was 59 percent. Causes of late death were arrhythmia, congestive heart failure, pulmonary hypertension, and myocardial infarction. For corrected transposition, Ilbawi et al.[15] have reported intermediate results in 12 patients undergoing anatomic correction. Operative mortality was 9 percent, the incidence of surgical atrioventricular block requiring pacemaker was 9 percent, and superior caval venous obstruction occurred in 9 percent of the patients. It was necessary to replace the conduit in 5 patients (45 percent) at a mean of 5 years. The investigators at Boston Children's Hospital have also recently summarized their experience with anatomic repair for congenitally corrected transposition.[16] They performed surgery in 44 patients between 1992 and 2005, using the Rastelli option in 23 and the double switch in 21. Early mortality was 4.5 percent. Left ventricular function, however, had deteriorated in almost one-fifth of the cohort after a median follow-up of only 2.7 years. This was particularly associated with the patients requiring a Rastelli procedure, and those who required a pacemaker.

In summary, therefore, it would appear that the operative mortality of anatomic repair is either slightly higher or similar to that of classical repair. There will be a need to replace conduits in all patients in whom they have been inserted, and left ventricular dysfunction at intermediate follow-up is already seen in one-fifth. Atrial arrhythmias must be anticipated in at least half at 10 years, and the incidence of baffle obstructions may be from 10 percent to 15 percent after 10

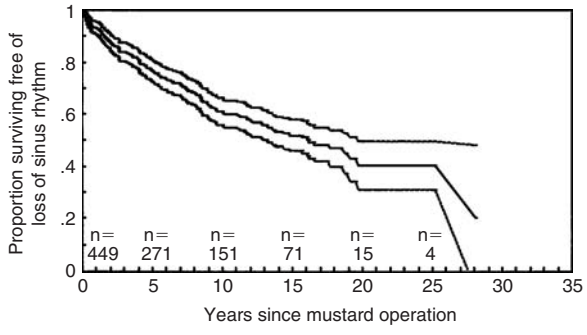

Figure 1.
Kaplan-Meier estimates of late survival free of loss of sinus rhythm after the Mustard procedure. Top and bottom lines indicate 95% confidence intervals. (Reprinted with permission from the American College of Cardiology Foundation.)

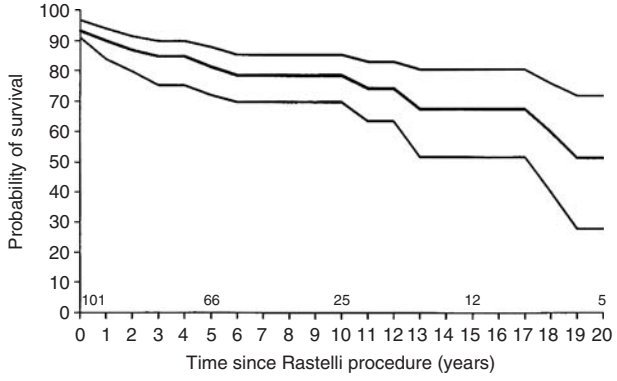

Figure 2.
Overall survival following Rastelli repair with 95% confidence limits out to 20 years. (Reprinted with permission from the American Association for Thoracic Surgery.)

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years. These patients are also at risk for coronary arterial stenosis. Long-term follow-up of patients undergoing anatomic repair is just now becoming available, and it will prove interesting to compare the results to the known long term outcomes of classical repair.

One-and-a half ventricular repair

We initially began utilizing this option for repair because neither the classical nor anatomic alternatives seemed optimal for these patients.[7] The idea behind the repair is that, in many of these patients, a pulmonary valvotomy alone is inadequate to provide an acceptable pressure in the morphologically left ventricle. The ideal left ventricular pressure in these patients, however, is different than in, for example, repair of tetralogy of Fallot, where one would aim for a low pressure in the subpulmonary ventricle. The goal in a patient with congenitally corrected transposition is to keep the pressure sufficiently high to maintain the septum in the midline. This midline location will improve morphologically right ventricular function, and decrease the amount of morphologically tricuspid insufficiency. The bidirectional Glenn, or superior cavopulmonary, anastomosis will unload the morphologically left ventricle sufficiently so that, despite the residual pulmonary stenosis, the left ventricular pressure will be acceptable.

The importance of septal location has been known for some time. Patients who have had a prior atrial baffle procedure for simple transposition have been treated by banding the pulmonary trunk in preparation for eventual conversion to an arterial switch. This experience revealed that, in many cases, as the left ventricle expanded in size with the increased afterload from banding, the septum shifted back to the midline.[17,18] This was associated with a striking decrease in the degree of tricuspid valvar insufficiency, and an improvement in right ventricular function.

Another advantage of the one-and-a-half ventricular option for the select patient is that it will avoid the placement of a conduit from the left ventricle to the pulmonary arteries. Placement of conduits frequently means that the tube crosses the midline, with the potential that it may be compressed by the sternum, leading to early failure.[19] It can also cause coronary arterial compression, leading to myocardial ischaemia and ventricular dysfunction. Changes of conduit are particularly dangerous when they cross beneath the sternotomy incision.[20] Avoidance of a conduit, therefore, is a distinct advantage of the one-and-a half ventricular approach. A further problem with the conduit in classical repairs is that, since it unloads completely the left ventricle, it allows the septum to shift too far into the left ventricular cavity, splaying open the morphologically right atrioventricular valve,

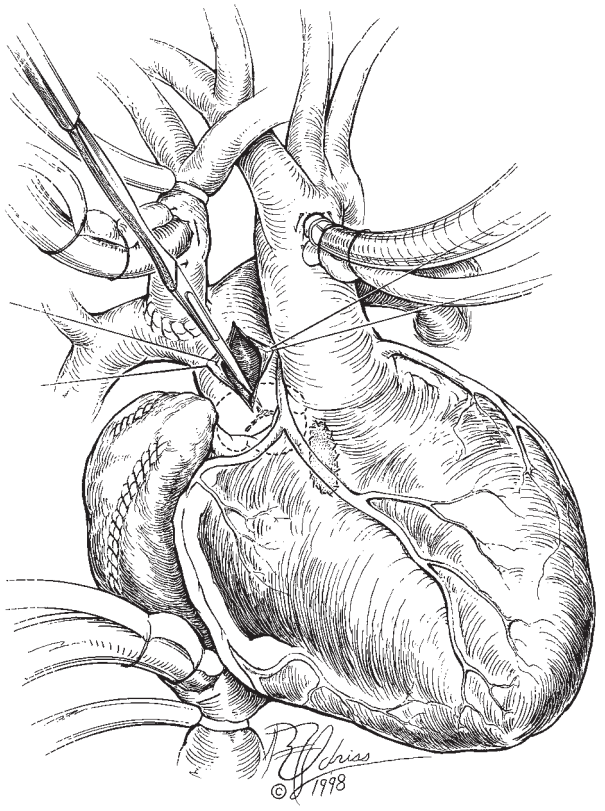

Figure 3.
Diagrammatic representation of a pulmonary valvotomy being performed through the pulmonary trunk in a patient with congenitally corrected transposition, ventricular septal defect, and pulmonary stenosis. The bidirectional Glenn anastomosis has been completed, and the ventricular septal defect closed through the right atrium.

and leading to valvar insufficiency and a progressive feedback loop of greater insufficiency and worsening ventricular function. We have now employed the one-and-a-half ventricular option in 7 patients.[21] Their mean age at the time of surgery was 5.8 plus or minus 4.5 years, with a range from 6 months to 11.5 years. Of the patients, 3 had co-existing ventricular septal defects and pulmonary stenosis, while 4 had isolated pulmonary stenosis. Previous operations had included construction of a modified Blalock-Taussig shunt in 2 patients, and pulmonary valvotomy and pulmonary arterioplasty in 1 patient each. Our objective was to close the ventricular septal defect, and relieve obstruction across the morphologically left ventricular outflow tract by subpulmonary resection and/or valvotomy (Figs. 3 and 4). We unloaded the volume on the morphologically left ventricle by the bidirectional Glenn so as to lower the left ventricular pressure, aiming to achieve a ratio to morphologically right ventricular pressure of 0.5 to 0.75. There was no early or late mortality, and there were no episodes of surgical atrioventricular block. The systolic left ventricular pressure measured preoperatively by cardiac catheterization was a mean of 98 millimetres of mercury.

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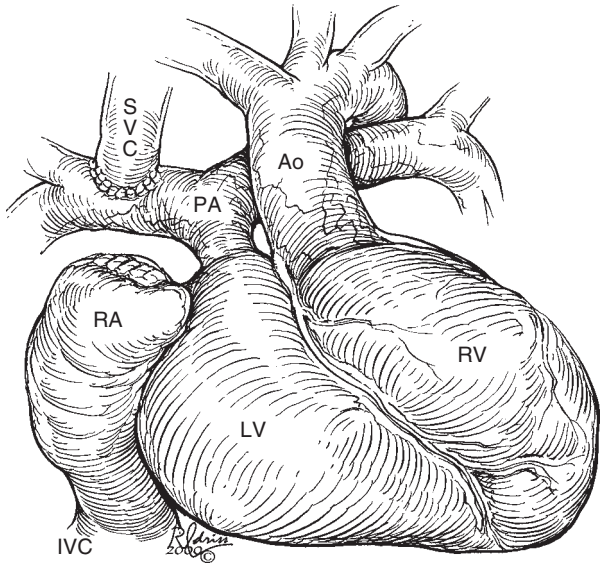

Figure 4.
The completed one-and-a half ventricle repair. Abbreviations: Ao: aorta; IVC: inferior caval vein; LV: left ventricle; PA: pulmonary trunk; RA: right atrium; RV: right ventricle; SVC: superior caval vein.

Following the one-and-a-half ventricular repair, this had fallen to 60 millimetres of mercury, with the mean pressure in the superior caval vein measured at 13 millimetres of mercury. Reoperation was needed in one patient for recurrent pulmonary stenosis. All patients are currently doing well at a mean follow-up of 33 months, with a range from 4 to 66 months.

Conclusions

Classical repair of patients with congenitally corrected transposition leaves the morphologically right ventricle fulfilling a systemic role, and consists of operative repair of the associated lesions, such as ventricular septal defect, pulmonary stenosis, morphologically tricuspid regurgitation, and third degree atrioventricular block. The mortality of these interventions is quite low, albeit that there is a known poor, but not completely unacceptable, long-term outcome. When combined with a bidirectional Glenn procedure to produce a one-and-a-half ventricular option, classical repair can be very efficacious in suitably selected patients, since not only does the procedure maintain the septum in the midline, improving right ventricular and tricuspid valvar function, but it can also avoid placement of a conduit from the left ventricle to the pulmonary arteries. Although anatomic repair, with either a double switch procedure or an atrial redirection combined with the Rastelli procedure, is becoming increasingly popular, the long-term complications remain unknown. These options include two operations that are known to harbour significant complications over the long term, leaving the question "do two wrongs make a right?"

References

1. Termignon JL, Leca F, Vouhe PR, et al. "Classic" repair of congenitally corrected transposition and ventricular septal defect. Ann Thorac Surg 1996; 62: 199–206.
2. Yeh T Jr, Connelly MS, Coles JG, et al. Atrioventricular discordance: results of repair in 127 patients. J Thorac Cardiovasc Surg 1999; 117: 1190–1203.
3. Biliciler-Denktas G, Feldt RH, Connoly HM, Weaver AL, Puga FJ, Danielson GK. Early and late results of operations for defects associated with corrected transposition and other anomalies with atrioventricular discordance in a pediatric population. J Thorac Cardiovasc Surg 2001; 122: 234–241.
4. Rutledge JM, Nihill MR, Fraser CD, Smith OE, McMahon CJ, Bezold LI. Outcome of 121 patients with congenitally corrected transposition of the great arteries. Pediatr Cardiol 2002; 23: 137–145.
5. Ilbawi MN, DeLeon SY, Backer CL, et al. An alternative approach to the surgical management of physiologically corrected transposition with ventricular septal defect and pulmonary stenosis or atresia. J Thorac Cardiovasc Surg 1990; 100: 410–415.
6. Yamagishi M, Imai Y, Hoshino S, et al. Anatomic correction of atrioventricular discordance. J Thorac Cardiovasc Surg 1993; 105: 1067–1076.
7. Mavroudis C, Backer CL, Kohr LM, et al. Bidirectional Glenn shunt in association with congenital heart repairs: the 1(1/2) ventricular repair. Ann Thorac Surg 1999; 68: 976–982.
8. Delius RE, Rademecker MA, de Leval MR, Elliot MJ, Stark J. Is a high-risk biventricular repair always preferable to conversion to a single ventricle repair? J Thorac Cardiovasc Surg 1996; 112: 1561–1568.
9. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. Br Heart J 1995; 74: 57–59.
10. Connelly MS, Liu PP, Williams WG, Webb GD, Robertson P, McLaughlin PR. Congenitally corrected transposition of the great arteries in the adult: functional status and complications. J Am Coll Cardiol 1996; 27: 1238–1243.
11. Nieminen HP, Jokinen EV, Sairanen HI. Late results of pediatric cardiac surgery in Finland: a population-based study with 96% follow-up. Circulation 2001; 104: 570–575.
12. Gelatt M, Hamilton RM, McCrindle BW, et al. Arrhythmia and mortality after the Mustard procedure: a 30-year single-center experience. J Am Coll Cardiol 1997; 29: 194–201.
13. Kreutzer C, De Vive J, Oppido G, et al. Twenty-five-year experience with Rastelli repair for transposition of the great arteries. J Thorac Cardiovasc Surg 2000; 120: 211–223.
14. Dearani JA, Danielson GK, Puga FJ, Mair DD, Schleck CD. Late results of the Rastelli operation for transposition of the great arteries. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2001; 4: 3–15.
15. Ilbawi MN, Ocampo CB, Allen BS, Barth MJ, Roberson DA, Chiemmongkoltip P, Arcilla RA. Intermediate results of the anatomic repair for congenitally corrected transposition. Ann Thorac Surg 2002; 73: 594–599.
16. Bautista-Hernandez V, Marx GR, Gauvreau K, Mayer Jr JE, del Nido PJ. J. Maxwell Chamberlain memorial paper for congenital heart surgery: determinants of left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries. Presented at 42nd annual meeting, Society of Thoracic Surgeons, Chicago, Illinois. January 30, 2006.

17. van Son JA, Reddy VM, Silverman NH, Hanley FL. Regression of Tricuspid Regurgitation after two-stage arterial switch operation for failing systemic ventricle after atrial inversion operation. J Thorac Cardiovasc Surg 1996; 111: 342–347.
18. Mavroudis C, Backer CL. Arterial switch after failed atrial baffle procedures for transposition of the great arteries. Ann Thorac Surg 2000; 69: 851–857.
19. Dunn JM, Start J, de Leval M. Avoiding compression of extracardiac valved conduits. Pediatr Cardiol 1983; 4: 235–238.
20. Stark J. The use of valved conduits in pediatric cardiac surgery. Pediatr Cardiol 1998; 19: 282–288.
21. Mavroudis C, Backer CL. Physiologic versus anatomic repair of congenitally corrected transposition of the great arteries. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2003; 6: 16–26.

https://doi.org/10.1017/S1047951106000801 Published online by Cambridge University Press