Surgical correction of scoliosis in Rett syndrome: cord monitoring and complications

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Abstract

Aim Rett syndrome is a progressive neurodevelopmental disorder that predominantly affects females and is associated with a high incidence of scoliosis and epilepsy. There is scant published work about intraoperative spinal cord monitoring in these patients and little more regarding the rate of perioperative complications. We investigated our institutions' experience with both.

Methods We retrospectively reviewed the records of 11 patients with Rett syndrome who underwent surgical correction of scoliosis at our institution between 2004 and 2010.

Results Eleven patients underwent successful correction of their scoliosis at an average age of 12. Eight of the patients suffered one or more significant complications. The average curve was corrected from 71° to 27°. Successful spinal cord monitoring was achieved in eight of the nine patients where it was attempted. No patient suffered any neurological complications. Average inpatient stay was 18.2 days.

Conclusion Scoliosis surgery in patients with Rett syndrome carries a very high rate of complications and an average hospital stay approaching 3 weeks. Both caregivers and surgeons should be aware of this when planning any intervention. These patients frequently have useful lower limb function and spinal cord monitoring is a valid tool to

aid in its preservation. We would suggest aggressive optimisation of these patients prior to surgery, with an emphasis on nutrition.

Keywords Scoliosis · Rett · Monitoring

Introduction

Rett syndrome (RTT) is a progressive neurodevelopmental disorder, first described by Dr Andreas Rett in 1966 [1]. It was largely overlooked until Hagberg presented a case series of 35 patients in 1983 [2] and gave the syndrome its eponym. It is a mostly sporadic sex-linked disease affecting approximately 1 in 10,000 [3] females, and is caused by mutation of the MECP-2 gene (methyl CpG binding protein 2), located on the long arm of the X chromosome [4]. Males with a mutated MECP-2 gene almost invariably do not survive to term, and those that do will die before the age of 2 from severe encephalopathy.

The syndrome is notable for its distinctive time course and pattern of involvement. It is characterised by four stages. The antenatal period is unremarkable and the child develops normally for approximately the first 6 months of life, at which point development slows. The second stage occurs between 1.5 and 3 years of age and is characterised by regression of acquired volitional hand movements and speech. There can be superimposed social withdrawal or irritability that may resemble autism and affected children will also go on to develop stereotypical hand movements, epilepsy and difficulty mobilising. This regression stage is succeeded by a plateau phase where symptoms stabilise for several years until the eventual progression to the late motor deterioration stage notable for scoliosis, progressive dystonia, rigidity and worsening ambulation.

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Interestingly, there is no gross abnormality found on neuropathological examination of the brain in patients with RTT [5], although overall brain size is reduced and neurons more tightly packed. Crucially, there is no evidence of degeneration or atrophy, defining Rett syndrome as a neurodevelopmental rather than neurodegenerative disorder. Approximately 60 % of patients will live to be 50 [6], though true longevity is unknown.

The overall incidence of scoliosis in Rett syndrome is estimated at around 64 % [7] and the general consensus is that curves should be treated when greater than 40°–50° [8] to improve sitting balance, perceived pain and ease of activities of daily living [9]. However, up to 100 % of patients will suffer some form of complication [10] and thus RTT patients present a considerable problem to the surgical team. These patients are typically underweight and undernourished [11], which combined with poor mobility results in osteopaenia/porosis with potential implications for the integrity of any bone/implant interface.

We chose to examine our experience with Rett syndrome patients and look in particular at the risk of perioperative complications and our ability to monitor the spinal cord during surgical correction.

Methods

We performed a retrospective chart review on 11 patients who underwent correction of scoliosis at our institution between 2005 and 2011. All patients had at least 2 years of follow-up at the time of writing.

Results

Eleven patients underwent surgical correction of scoliosis at an average age of 12 years and 7 months (range 7–20). The majority of the curves were long, neuromuscular, C-shaped curves, but one patient had a double major curve and one an isolated thoracic curve. No patient underwent formal flexion or traction films preoperatively. The mean major curve Cobb angle measured 70.7° (range 44–105).

All patients underwent posterior instrumented fusion of their curves without any form of anterior release. The instrumentation used progressed from hybrid constructs utilising pedicle screws and sublaminar wires to the more modern all-pedicle-screw construct. Eight of the 11 patients were noted to have some form of complication during the course of their inpatient stay (Table 1). The length of inpatient stay varied from 8 to 42 days (mean 18.2).

Eight of the patients were defined as ‘mobile’—which by the limited status found in RTT we have defined as ‘able

Table 1 Complications and length of stay

Patient	Complications	Inpatient stay (days)
1	Superficial wound infection, persistent diarrhoea	23
2	Haemothorax, extended intubation, superficial wound infection, UTI sepsis, aspiration pneumonia	42
3	None	14
4	None	8
5	Deep wound infection requiring washout and prolonged antibiotics	31
6	None	9
7	Severe hypokalaemia	9
8	Pneumothorax, hospital-acquired pneumonia	16
9	Superficial wound infection	15
10	UTI, aspiration pneumonia.	13
11	Hospital-acquired pneumonia	20

to ambulate, stand or crawl, with or without assistance or aids’. This generous description was necessitated by the disabled nature of this patient cohort. Those patients who had observable function in terms of mobility or continence had monitoring attempted (7 of the 11) with sensory evoked potentials (SEPs), and a further two with SEPs and motor evoked potentials (MEPs). In eight of the patients, consistent reliable monitoring was achieved from the outset, and in the last patient monitoring had to be abandoned. One patient was noted to have variable SEPs that were associated with episodes of hypotension rather than corrective manoeuvre. The SEPs in this patient remained variable throughout the procedure and no wake-up test was undertaken. No patient was found to have a neurological deficit postoperatively (Table 2).

The size of curves and final postoperative results are noted in Table 3. The average residual curve was 27°. No patient had significant deterioration in their curve, failure of construct, or failure of bone/implant interface during follow-up (mean 5 years, range 2–8).

Discussion

In common with most patients with a syndromic scoliosis, RTT patients run a considerable risk of complications [9]. Most of the complications we encountered were infective—four pneumonias, four wound infections and two urinary infections. This had considerable implications on the length of hospital stay: patients without complications were discharged an average of 8.7 days postoperatively, whereas those with complications remained as inpatients for an average of 21.8 days.

Table 2 Preoperative function and spinal cord monitoring

Patient	Preoperative function	Spinal cord monitoring
1	Able to stand if assisted	Variable SEP responses during surgery
2	Able to stand with an aid	SEPs stable throughout
3	No useful lower limb function	No monitoring used
4	Able to walk a few steps	SEPs stable throughout
5	No useful lower limb function	Unable to achieve good baseline—abandoned
6	Able to walk a few steps	SEPs stable throughout
7	Able to roll herself over	SEPs stable throughout
8	Able to walk with a frame	SEPs stable throughout
9	Able to walk with a frame	SEPs and MEPs stable throughout
10	Able to walk a few steps	SEPs and MEPs stable throughout
11	No useful lower limb function	No monitoring used

Table 3 Cobb angles and pelvic obliquity

Patient	Cobb angle		Pelvic obliquity	
	Preop	Postop	Preop	Postop
1	74	28	27	Level
2	50	15	Level	Level
3	72	32	28	10
4	72	29	Level	Level
5	70	46	45	28
6	65	21	Level	Level
7	70	23	28	13
8	105	33	22	11
9	54/63	18/23	Level	Level
10	101	33	40	6
11	44	10	13	Level

It is perhaps not surprising that the degeneration of scoliosis and the nutritional decline of the patient frequently coincide. RTT patients are known to become progressively underweight as they age, and scoliosis generally progresses during the late motor deterioration phase which is associated with poor feeding. Previous work has established that early gastrostomy formation reverses some of the weight loss observed in RTT patients [10] and it would seem prudent that these patients receive extensive optimisation before surgery. Decisions regarding intervention should therefore be made early to allow involvement of a multi-disciplinary team including paediatric surgical, dietetic and gastroenterology

teams to ensure that these patients are considered for supplemental feeding. If susceptibility to complications can be reduced, there will be obvious consequences in terms of reduced morbidity and shorter hospital stays.

With regard to curves, we were able to achieve a good correction in the vast majority of cases. Despite the lack of bending films, the patients were clinically flexible and the two curves originally in excess of 100° corrected down to around 33°. All except one patient had significant correction of pelvic obliquity. The exception was the oldest of all the patients at 21 years and fully skeletally mature. As one might expect, her deformity was significantly more difficult to correct than the younger patients and this serves to highlight the importance of intervening prior to the completion of skeletal maturation. It is reassuring to note that even large curves remain flexible and correct well with posterior-only intervention.

Despite the poor bone quality of the RTT patients, there was no observed screw pullout or construct failure noted throughout the period of follow-up.

Whilst it is well established that monitoring is possible in the majority of syndromic scoliosis patients [11], it is not universally utilised. In this limited series, the use of spinal cord monitoring followed a predictable course—wherein all of those patients in whom there was function to be preserved had an attempt to monitor spinal cord function intraoperatively. We were successful in the majority and utilised SEPs in all cases. MEPs were usually avoided due to concerns over their use in patients with epilepsy. This concern does not appear to be well founded and so MEPs have now become part of routine monitoring in these cases.

Conclusion

Scoliosis in Rett patients is fraught with risks, in our institution's experience, which certainly tallies with other published data [9]. Complications, which remain the norm rather than the exception, lengthen inpatient stay dramatically. These patients frequently have distal functioning that we would aim to preserve and spinal cord monitoring is a feasible method of achieving this. It should therefore be considered as part of routine management in all patients. Whilst it could be acceptable to operate on a small number of select cases without the benefit of cord monitoring, these should remain the minority.

Conflict of interest None.

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