

CORRECTIVE SCOLIOSIS SURGERY IN SMITH MAGENIS SYNDROME: A CASE REPORT

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INTRODUCTION:

Smith–Magenis Syndrome (SMS) is a complex and rare multisystem congenital condition, with 30% of cases associated with scoliosis.¹ Distinguished anatomical components, such as dysplastic pedicle, the extent of deformity, intraoperative blood loss and fusion efficacy are part of the challenges faced throughout the care of these patients.

REPORT AND DISCUSSION:

This is a case of a 14-year-old, Malay girl with underlying heart anomalies and autism, presented to us after scoliosis screening at school. Initial brace treatment was unsuccessful, as she developed severe progressive thoracic scoliosis, with radiographs demonstrating rigid curves and a Cobb's angle measuring 85° (Figure 1). In addition, Computed Topography (CT) scan was done to assess bone morphology. Successfully obtaining clearance for surgery from pediatricians and cardiologists, she underwent posterior spinal instrumentation, supplemented by autologous graft and demineralized bone matrix. Uneventful post-operatively, she was hemodynamically stable with no signs of a wound infection and was discharged home well 3 days post-surgery. Good correction of scoliotic curve up to 15° and a balanced spine in both coronal and sagittal planes was achieved (Figure 2). Follow-up to 4 months post-surgery showed a well healed wound, with no loss of deformity correction, and a good clinical outcome. Pre-operative imaging act as a template to analyze anatomical configuration, thus reducing the risk of pedicle screw perforation. Furthermore, the use of cell-saver is crucial in reducing intra-operative blood loss, whilst additional allograft improves fusion efficacy by providing an osteoinductive and osteoconductive environment for bone growth.



Figure 1: Pre-operative Radiographs

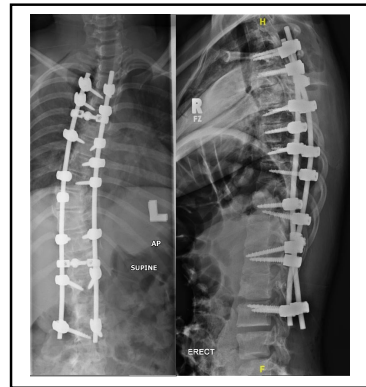


Figure 2: Post-operative Radiographs

CONCLUSION:

Despite the challenges, a good team with multidisciplinary approach, thorough pre-operative planning is vital, in order to perform safe surgery, achieve good outcome and avoiding post-operative complications on this group of patients.

REFERENCES:

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