Complications of Occipitocervical Fusion in Children with Skeletal Dysplasia
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INTRODUCTION: Cervical instability is a consistent orthopaedic manifestation in children with skeletal dysplasias. The occipitocervical complex sometimes needs to be stabilized with an occipitocervical arthrodesis in order to address instability, deformity or myelopathy. It is assumed that due to their skeletal abnormality, these children will have a high complication rate both perioperatively and postoperatively, but the literature is sparse on this subject. The purpose of this study was to retrospectively review our experience with the surgical management of occipitocervical instability in children with skeletal dysplasia. Specifically, we attempted to report the complication rate of occipitocervical fusion in this patient population in both the perioperative and postoperative periods.

METHODS: Twenty-one children previously diagnosed with a skeletal dysplasia underwent a posterior occipitocervical fusion for instability at our institution from 1980 to 1998. A retrospective review was performed in order to determine the complications from this procedure.

RESULTS: Of the 21 children with skeletal dysplasia that underwent an occipitocervical fusion for instability, 4 were excluded from the study because they had prior procedures done elsewhere. Of the remaining 17 patients, 8 were male, and 9 were female. Five patients had congenital spondyloepiphyseal dysplasia, 4 had metatropic dysplasia, 2 had spondyloepimetafysyal dysplasia, and 1 each had spondylometaphyseal dysplasia, camptomelic dysplasia, achondroplasia, metaphyseal chondrodysplasia, pseudochondroplastic dysplasia, and Dyggve-Melchior-Clausen syndrome. Five of the patients exhibited myelopathy of varying severity. At a mean age of 6 yrs. + 2 mo., 17 children underwent an occipitocervical fusion with bone graft. Eight children had only autograft bone, 8 children had a mixture of auto and allograft bone, and 1 child had only allograft bone; there was no instrumentation used in any of the patients. Postoperatively, 16 patients were immobilized in a halo cast and 1 in a hard collar; the immobilization period averaged 13 weeks.

The mean follow up period was 40 months. Two children had recurrent instability due to pseudarthrosis and underwent a revision posterior cervical fusion. Both children were immobilized in a halo vest after the index procedure.
for a mean of 10 weeks; both had allograft bone graft. Another child developed
junctional degeneration and instability necessitating an extension of her fusion
mass subaxially about 10 years later. Three of the 5 myelopathic children had no
improvement neurologically. Eleven of the 16 children immobilized in halos
developed pin tract complications. Additionally, there were 3 children with
pulmonary problems, which prolonged their hospitalization, 2 falls necessitating
early halo removal, one patient had a transient CN IV palsy, and one patient
with a superficial wound infection.

DISCUSSION/CONCLUSION: The surgical management of cervical
instability in children with skeletal dysplasia, although rare, is complicated.
Many of the skeletal dysplasias manifest rapidly progressive deformities and/or
instability that can be lethal. Occipitocervical arthrodesis in these children can
stabilize the pediatric spine, but the outcome may not be similar to the
otherwise normal child with a traumatic or idiopathic pediatric spine deformity.
Two of the 17 children (12%) in our study had a pseudarthrosis and recurrent
instability. In addition to those two children, another child needed revision
surgery to fuse a junctional level degeneration. Furthermore, the prognosis for
neurologic improvement in the myelopathic children was guarded. Overall, the
complication rate (major and minor) was 76%. The prolonged use of the halo
cast in patients with skeletal dysplasia is almost always required for
postoperative immobilization, but comes with a high rate (69%) of pin site
problems. To limit complications in this difficult group of patients, we
recommend a meticulous bony decortication, fusion with autograft bone and
halo cast immobilization with excellent pin care.