CERVICAL MYELOPATHY SECONDARY TO ATLANTOAXIAL INSTABILITY IN DOWN SYNDROME: A CARE REPORT

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Introduction: Os odontoideum in down syndrome associated with atlantoaxial instability is more often presented in childhood or adolescence. Most of all authors are recommend early fusion if present of atlantoaxial instability and for all patients with Os Odontoideum. This is a report of a Down syndrome child with spontaneous atlantoaxial subluxation causing spinal cord compression.

Discussion: 7 years old girl with underlying down syndrome, presented with gradual worsening of bilateral upper limb weakness associated with difficulty to ambulate and urinary incontinence for 2 weeks prior to admission. Her neck kept in flexed and her head tilted to the left side with reduced neck range of motion. Neurology examination bilateral upper and lower limb was grade 2-3 with upper motor neuron lesion feature. She also had cervical myelopathy signs. The plain radiograph and CT cervical showed subluxation of C1/C2 with atlanto-dens interval (ADI) 9.7 mm and space available for cord (SAC) 5.3 mm. Presence of Os odontoideum (dystrophic type) with C1/C2 subluxation causing spinal canal stenosis and spinal cord oedema on MRI. She underwent posterior fusion of C1C2 with two separate iliac autografts are placed between C1 and C2 and wrapped with ethibond suture 5/0 as a sublaminar suture. Halo-vest was applied following the wound closure. The observation period was 1 year. She had a superficial pin site infection at two months post-op and treated with antibiotics only. The neurology was improved, from grade 3 to grade 4-5 on both upper and lower limbs, patient able to ambulate with support, and normal micturition. MJOA score 3 pre-operatively and 11 post-operatively.

Conclusion: Autologous graft C1/C2 fusion with the sublaminar suture is generally feasible in challenging atlantoaxial instability. Various complications associated with the surgery should be informed to the parent and extensive evaluation pre-operatively to avoid various complications.