IMPLANTABLE DIRECT CURRENT STIMULATION OF CERVICAL FUSION IN A PATIENT WITH HAJDU-CHENEY SYNDROME

Alan P. Moelleken, M.D.,
514 West Pueblo Street
Santa Barbara, CA 93105

INTRODUCTION

Hajdu-Cheney syndrome, a rare congenital disease, is characterized by acro-osteolysis with diffuse osteoporosis in the absence of other associated diseases.[1] The proposed mechanism by which this occurs has been thought to be reduced bone formation. However, radius bone biopsy has revealed both reduced bone formation and increased osteoclastic bone resorption.[2] This suggests that the osteolysis and trabecular osteoporosis are produced by the same mechanism.[3] The radiographic and clinical sequelae characteristic of this syndrome develop and progress with age.[4] Implantable direct current (DC) stimulation has been used in the lumbar spine since the late 1980's and has been shown to benefit spine fusion patients, particularly those considered difficult to fuse. The beneficial effect of DC stimulation on bone graft with or without instrumentation has been well documented in clinical investigations.[5-10]

Figure 1


Figure 2


Figure 3


Figure 4A


Figure 4B

Figure 5A

CASE PRESENTATION

Patient FK, a 62-year old male, presented with severe myelopathic symptoms due to cervical spine instability with subluxation at C3-4 and C7-T1, secondary to acro-osteolysis and osteoporosis (Figure 1). Further radiographic, clinical, and neurological examination revealed phalangeal, skull and other abnormalities consistent with Hadju-Chaney Syndrome. Cervical myelopathy was progressing rapidly, and halo brace stabilization by a different surgeon had failed due to the poor quality of the patient's bone with gross loosening of the cranial pins (Figure 2). He was subsequently placed in a Minerva Cervical Brace (USMC) with further disease progression and felt to be "inoperable" due to his severe osteoporosis.


Figure 5B

METHODS

Under our care surgical treatment consisted of partial anterior vertebectomy and interbody fusion at C3-4 and C7-T1 using autogenous iliac crest bone graft. Posterior cervical fusion was then performed from C2 through T1 with interspinous wiring. Since consolidation of the posterior graft would be crucial to the success of the construct a direct current stimulator with the cathodes in contact with the posterior bone graft was implanted. Interspinous wiring was performed using the available spinous processes with adequate bone quality (Figure 3).

Multiple abnormalities were demonstrated intraoperatively. All vertebrae appeared abnormal with multiple areas of laminae absent, particularly C4, where dura was present with no overlaying lamina. The spinous processes of C3 and C4 were completely absent and other spinous processes were deficient.

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RESULTS

Postoperatively, the patient was unchanged neurologically and placed in a Minerva Cervical Brace for 6 months. Progression of myelopathic symptoms were ceased and neurological improvement was noted (Figure 4a and 4b). The patient is now 10 years post-op with a solid fusion (Figure 5a and 5b).

CONCLUSIONS

The procedure performed, while high-risk, did stop the progression of his myelopathy and progressive deformity. The implantable stimulator may have helped considerably in obtaining a solid fusion in this difficult situation.

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