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Spinal correction in patients with Fukuyama congenital muscular dystrophy.

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Abstract

BACKGROUND: Fukuyama congenital muscular dystrophy (FCMD) is one of the most common congenital progressive muscular dystrophies in Japan. Some patients develop a severe spinal deformity that leads to an unstable sitting position or pain. Since 2008, we have treated FCMD using posterior spinal fusion. This study reports the short-term clinical and radiographic results of posterior spinal correction and fusion in FCMD.

METHODS: We retrospectively reviewed 11 consecutive FCMD patients, average age 13 years old, treated with posterior spinal instrumentation and fusion between 2008 and 2015. All patients were non-ambulatory and complained about difficulty sitting and/or buttock pain. Posterior spinal correction was performed to halt progression of spinal deformity and improve their sitting balance. Assessment was performed clinically and with radiological measurements at a mean follow up period of 34.5 months. To evaluate functional status of patients after surgery objectively, a Muscular Dystrophy Spine Questionnaire (MDSQ) was obtained at the final follow up.

RESULTS: The mean height, weight and body mass index of the patients were 144.1 ± 11.8 cm, 26.5 ± 8.7 kg and 12.5 ± 2.6 kg/m². The average intensive care unit stay was 1.3 days. Five patients had complications related to surgery. The mean preoperative major Cobb angle and pelvic obliquity (PO) were $65.5 \pm 41.7^\circ$ and $31.4 \pm 28.9^\circ$, respectively, were corrected to $34.4 \pm 29.7^\circ$ and $20.0 \pm 18.7^\circ$ just after the surgery, and were maintained at $35.1 \pm 29.9^\circ$ and $20.5 \pm 21.1^\circ$ at the final follow up. The average MDSQ score was 35.8 ± 13.2 at the final follow up.

CONCLUSION: Posterior spinal correction and fusion in FCMD achieved good radiographic results and clinical improvement with acceptable perioperative complications. FCMD patients are mentally impaired and physically small, so post-operative observation and close attention to perioperative complications are critical.

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