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Anaesthetic management of a paediatric patient with congenital fibre type disproportion myopathy.

[Article in English, Spanish]

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Abstract

Congenital fibre type disproportion (CFTD) is a rare type of myopathy that is characterised by muscle weakness and hypotonia during childhood. Clinical features include motor delay, feeding difficulties, limb weakness, joint contractures, and scoliosis. A report is presented of the anaesthetic management of a 3-year-old girl with CFTD myopathy associated with a mutation of the TPM3 gene, scheduled for adenotonsillectomy because of obstructive sleep apnoea hypopnoea syndrome (OSAHS). The main concerns were the possible susceptibility to malignant hyperthermia, the risk of anaesthesia-induced rhabdomyolysis, a greater sensitivity to non-depolarising muscle relaxants, and the presence of OSAHS. Total intravenous anaesthesia with propofol and the use of rocuronium/sugammadex appear to be safe options. Given the high risk of respiratory compromise and other complications, patients should be closely monitored in the post-operative period.

KEYWORDS: Anestesia intravenosa; Apnea obstructiva del sueño; Congenital fibre type disproportion; Congenital structural myopathies; Desproporción congénita del tipo de fibras; Intravenous anaesthesia; Miopatías estructurales congénitas; Obstructive sleep apnoea hypopnoea syndrome; Rocuronio; Rocuronium; Sugammadex

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