題名:Hereditary gingival fibromatosis associated with periodontal disease: clinical diagnosis and treatment

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摘要:BACKGROUND: Rutherfurd syndrome is a rare genetic disorder that is primarily characterised by the classical triad of gingival fibromatosis, delayed tooth eruption and corneal dystrophy. Associated features of the condition include abnormally shaped teeth, mental retardation and aggressive behaviour. CASE REPORT: His Consultant Clinical Geneticist referred a 2-year-old boy to the Dept. of Paediatric Dentistry at Manchester Dental Hospital. The child and his father had a diagnosis of Rutherfurd Syndrome; a rare autosomal-dominant condition featuring corneal dystrophy, gingival hypertrophy, abnormally shaped teeth and delayed eruption. The only erupted teeth were 52, 71 and 81. The patient also suffers from features of Marfan syndrome, a condition he has inherited from his mother. CLINICAL MANAGEMENT: Preventive advice was provided and the patient was placed on regular review. By the age of 4 years, 52, 61, 72, 71 and 81 were the only teeth present clinically. Maxillary and mandibular dentures were provided, which he refused to wear. FOLLOW-UP: At age 8 years, 31 and 41 were erupted lingual to the primary incisors. No other permanent teeth had erupted. Radiographs revealed failure of eruption of multiple primary and permanent teeth. The second premolars had formed above the unerupted mandibular primary second molars. The patient continued to refuse any intervention. Treatment options discussed included overdentures and /or removal of primary teeth and exposure of permanent teeth. CONCLUSION: This case has presented a rare syndrome with two important dental

features, namely gingival hyperplasia and failure of eruption. The case has a long term follow up of 6 years and has a first time association to Marfan syndrome. It also highlights the importance of family history and how it can affect the attitude of the child towards dental treatment.