Talon Cusps Presenting in a Child with Alagille's Syndrome – A Case Report

Mahua Chatterjee*/ Carol Mason **

Talon cusp is a rare dental anomaly often associated with systemic conditions such as Rubenstein-Taybi Syndrome, Ellis-van Crevald Syndrome and Incontentia pigmenti achromians. It has not been previously reported as occurring in Alagille Syndrome.

Early diagnosis of talon cusps is crucial for the correct management and avoidance of complications. This case highlights the need for careful dental examination when treating children with syndromes, as new phenotypes that can have an effect on the dentition.

Keywords: *Alagille syndrome, talon cusp, children*
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INTRODUCTION

Alagille Syndrome was first reported in 1975 by Alagille *et al.*[1] The authors noted the association between intrahepatic cholectasis and pulmonary arterial stenosis. The syndrome is characterized by low birth weight, growth retardation, vertebral anomalies, cardiovascular and hepatic complications.[2] Twenty five percent of patients tend to die before the age of five due to the cardiovascular and hepatic complications, but the disorder is usually benign.[3,4] The exact inheritance pattern is yet to be completely understood but is thought to have an autosomal inheritance with reduced penetrance and variable expressivity.[4,5]

Facial features include a broad forehead, triangular face and prominent zygomatic arches. The eyes tend to be deep set and they tend to suffer from a range of ophthalmic problems. The nose tends to be long and have a bulbous tip.

This case report describes a nine-year old girl who has been diagnosed with Alagille Syndrome. During her dental management it was noted that she had lateral incisors with talon cusps. There are no previously reported cases of this association in the literature.

CASE REPORT

The child presented is the youngest of six children in an orthodox Jewish family, one other child having died from a heart defect. She was born at thirty-eight weeks weighing 3 lbs 1 oz. It was an uncomplicated vaginal birth. A heart defect was noted in the child soon after birth, and she was diagnosed with severe pulmonary valve stenosis. Due to the child vomiting from birth, investigations including a barium swallow, pH studies and an endoscopy were carried out. At twelve weeks of age she was diagnosed with gastro-oesophageal reflux. She was prescribed the relevant medication and a nasogastric tube was inserted.

Feeding was a problem. Her feeding history indicates that she was breast fed for three months but that this was difficult. At six weeks she started bottle-feeding, but she could only take an ounce before vomiting. By five months the child could drink clear fluids and soup but could not tolerate any solid food in her mouth. She demonstrated poor weight gain and was well below the third percentile. At eight months a gastrostomy was inserted and as a result the vomiting began to decrease. This was removed at four years of age and since then she improved and can now eat mashed food and bread.

In addition to the cardiac and feeding problems previously mentioned she also developed gross and fine motor coordination difficulties, global developmental delay and persistent glue ears (for which she had grommets inserted) and she was found to have Intrahepatic duct deficiency. Due to the combination of symptoms described previously, especially the atypical liver involvement and the pulmonary valve stenosis the child was diagnosed with Alagille Syndrome.

The initial referral of the child to the Maxillofacial and Dental Department was at five years of age by her ENT Consultant. The presenting complaint was the appearance of the upper primary central incisors. The upper left primary central incisor appeared to be intruded and the upper right was mobile. There was no obvious history of trauma to the teeth.

Intraoral examination showed the child to have poor oral

*Mahua Chatterjee BDS MFDSRCS, Senior house officer
**Carol Mason BDS FDSRCS, Consultant department of Pediatric Dentistry, Great Ormond Street hospital.

Send all correspondence to: Dr. Mahua Chatterjee, 40 Illingworth Way, Enfield, London, EN1 2PA, UK

Tel: + 020 7829 7866
Fax: +020 7829 8804

E-mail: mahua77@tiscali.co.uk

hygiene and the presence of heavy calculus and plaque. She had a full primary dentition with the exception that the lower left lateral incisor was missing; in addition she had all four first permanent molars and her lower permanent central incisors. There was gingival recession around these lower central incisors and it was noted that the child had an anterior open bite - both these features were attributed to constant sucking of a dummy, which the child had bought to the visit. There were no carious cavities. The child had no dentist and this was her first dental visit. She was eating a mainly liquid diet, which contained a lot of sugar, as she needed to increase her calorie intake. Tooth brushing was erratic and on average the patient was brushing her teeth once a day.

As she was due to have grommet insertion and adenoidectomy, it was decided to remove both upper primary central incisors at the same time to avoid complications. Oral hygiene and diet advice were given, and the parents were advised to try and break the dummy sucking habit.

The child was kept under review and regularly attended appointments with the hygienist. A couple of years later when the permanent incisors had erupted it was noted that both upper permanent lateral incisors had talon cusps (Figs 1 and 2). Both teeth were caries free but the oral hygiene was still noted to be poor. It was decided to continue regular reviews with oral hygiene instruction and fluoride application, with view to fissure sealants as soon as cooperation allowed.

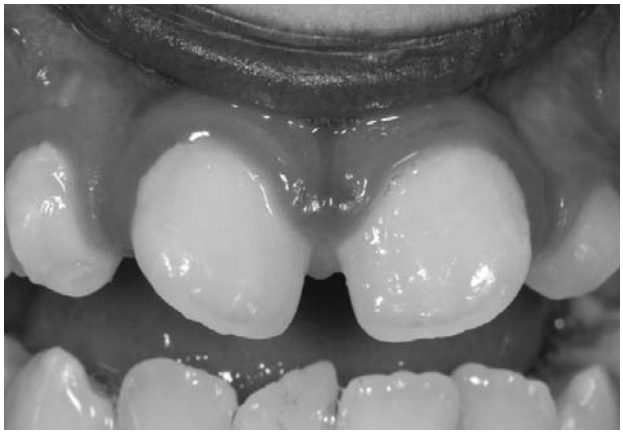

Figure 1. Photograph showing the labial aspect of the upper permanent incisors.

DISCUSSION

A talon cusp is a rare dental anomaly that manifests as an accessory cusp like structure in a tooth crown.[6] The shape of the cusp in this anomaly resembles an eagle's talon and hence has been given this name.[7] The cusp involves the palatal cingulum area of the permanent anterior teeth.[8] It is composed of enamel, dentin and occasionally presents a pulp horn.[9]

The cusp blends smoothly with the tooth, however there is a developmental groove in the area where the cusp joins the palatal surface of the tooth. This is an area of plaque retention and therefore is susceptible to carious attack.[10] Whether unerupted or partially erupted, the cusp can mimic a supernumerary tooth and this can lead to unnecessary surgical intervention. Teeth most often affected are the permanent maxillary incisors.[11]

The bulk of the cusp can cause occlusal disharmony but the degree to which this occurs depends upon the form of the talon cusp and this varies. The tip of the cusp may stand away from the rest of the crown increasing its labiopalatal width, and therefore the probability of occlusal interference. In patients where the cusp is in closer apposition to the palatal surface of the tooth the resultant fissure between the cusp and tooth often become carious. Also the tip of the cusp does not always coincide with the midline of the long axis of the tooth, and when deviation does occur it is often to the mesial aspect.[11]

Clinical problems associated with this defect include a compromised appearance of the tooth, occlusal interference, tooth displacement, caries and tongue irritation. It has also been reported that this condition can cause malocclusion and shift in the normal midline of the patient's dentition. The talon may fracture resulting in pulpal exposure and loss of vitality.

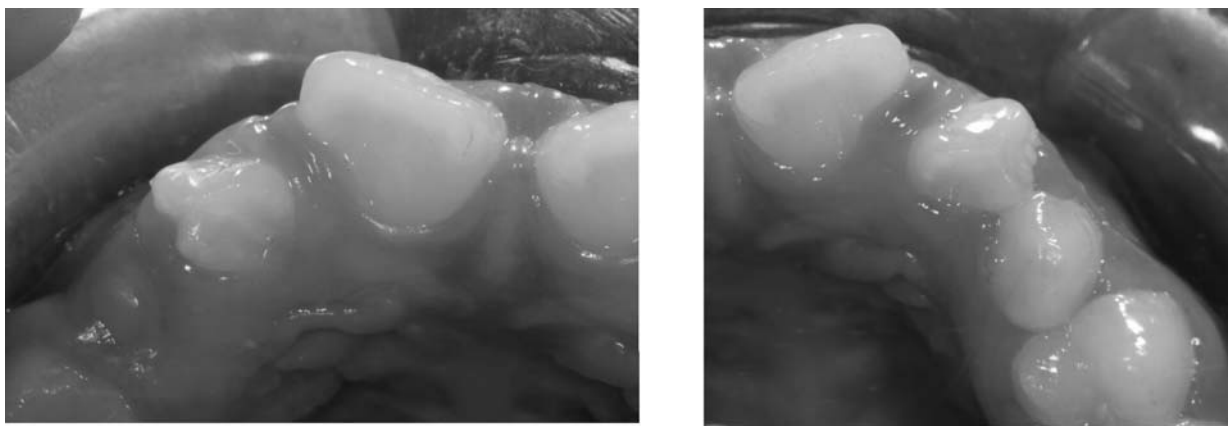

Figure 2. Photograph of the palatal aspect of the upper right and left lateral incisor.

A number of methods have been suggested in the treatment of talon cusps. Some have suggested that if occlusal accommodation is carried out that a pulpectomy must also be done as the cusp often involves pulpal tissue.[9] Other clinicians have reported simply grinding off the protruding cusp and then leaving the tooth in this state when no extension of pulpal tissue is found.[14] Gradual reduction of the cusp over a period of months, may allow reparative dentin formation and vitality to be retained.[15,16] Reduction of the cusp with a partial pulpotomy is also an option.

Talon cusps have been reported as occurring in conditions such as Rubenstein-Taybi Syndrome,[17] Ellis van Crevald syndrome,[18] Incontinentia pigmenti achromians.[19] As yet no case of talon cusps in a patient with Allagille Syndrome has been reported. Due to the factors discussed in the previous text it is important that talon cusps should be diagnosed early as its clinical features could influence future treatment.[11]

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