

7th Global Dentists and Pediatric Dentistry Annual Meeting

March 31- April 01, 2016 Valencia, Spain

Dental management for COACH syndrome

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Background: COACH is an acronym that outlines a syndrome –linked with: C - cerebellar vermis defect (Joubert syndrome), O– oligophrenia, A–ataxia, C–coloboma, H-hepatic fibrosis. Joubert Syndrome (JS) is characterized as a rare brain malformation by the absence or underdevelopment of the cerebellar vermis - an area of the brain that controls balance and coordination as well as a malformed brain stem in which the obligatory hallmark is the molar tooth sign through magnetic imaging.

Objective: This poster will present the oro-digital facial manifestation of JS and related disorders and present the experience in providing dental care.

Case reports: This poster reported two cases of Saudi pediatric male and female patients with growth retardation. Parents are first cousins supporting an autosomal recessive mode of inheritance. Male and female are equally affected and occurs in 1 to 100,000 live births. Optimal management requires a multidisciplinary approach with particular attention to feeding problems, ataxia and episode tachypnea. These factors should always be considered when dental treatment will be required for patient with JS. Decision was taken by group consultation for extraction of the anterior teeth under O2 supporting and LA. Two weeks later, tongue and lip healed with scar. Patient improved good sleeping habits and had stopped lip biting and improved psychologically.

Conclusion: It was concluded that early dental treatment for patients diagnosed with Joubert Syndrome is necessary to avoid possible future complications such as eating or feeding difficulties and dehydration.

Biography

Huda Salem Alrakaf has completed her Masters in Pediatric Dentistry in the year 1998 from King Saud University, Riyadh, Saudi Arabia. She also has obtained specialized certificate on High Risk Pediatric Dentistry (2002); as well as, completed her training in Pediatric Dentistry for Special Need Children (2003) from La Universidad Complutense de Madrid, Spain. She is at present a consultant in Pediatric Dentistry in Prince Sultan Medical City. Two of her publications are in the *International Journal for Pediatric Dentistry*.

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