Arkansas Children's
HOSPITALS · RESEARCH · FOUNDATION

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Health care professionals,

Arkansas Children's Hospital has been fortunate to host two International *SATB2*-associated syndrome (SAS) Conferences for children and their families, clinicians, and researchers. Dozens of individuals with SAS have been evaluated by several pediatric specialists including Genetics, Otolaryngology (ENT), Plastic surgery, Dental, Orthodontics, Neuropsychology, Speech Pathology, and Audiology.

Our Dental team has gathered data about children and young adults who have SAS through extensive clinical and radiographic evaluations. Our findings yielded the following information:

- In regards to dental characteristics, 85% presented with severe crowding and rotations. Other dental anomalies contributing to the crowding include anterior macrodontia and supernumerary teeth. Additionally, 28 of the patients were found to have malformed of anterior teeth.
- Over half of the patients evaluated had a skeletal class two malocclusion.
- Based upon analysis of radiographs from 23 individuals with SAS, 91% had very delayed or missing mandibular 2nd bicuspids, 30% had very delayed or missing maxillary 2nd bicuspids, and 17% were missing one more succedaneous mandibular anterior teeth. Our findings also indicated a significant delay in permanent molar root development.
- In regards to potential anterior and posterior pulp therapy, radiographic exposure is highly recommended as 22% had multiple pulp stones, either in incisors or molar teeth.
- Other common behavioral issues that can impact oral hygiene and dental care include a high tolerance to pain (97%), hyperphagia (72%), bruxism (62%), and balance abnormalities (75%).
- Due to issues maintaining balance, 56% of the children had anterior tooth trauma.
- Sialorrhea is reported in 85% of individuals but resolves in the majority of patients by 4-years of age.
- Feeding problems, specifically difficulty with chewing, overstuffing mouths, and pharyngeal dysphagia are common in children with SAS.

There is obvious individual variability among children who have SAS. Based on our findings from the 62 different children we were able to assess, our team recommends the following considerations for the patients' clinicians and educational team regarding dental care:

- As encouraged by the American Academy of Pediatric Dentistry, the first dental visit should be within the child's first year of life. We recommend seeing a pediatric dentist with special needs experience. Pediatric dentists often have hospital privileges and are able to treat the child under anesthesia if dental treatment is required.
- Bitewings or other radiographs can be recommended as deemed necessary based upon the child's caries risk and/or cooperation. Children with SAS have shown to display a range of dental cooperation from being compliant with in-office procedures to being resistant to regular check-ups. Frequent dental visits to desensitize these children are important.

- The first dental panoramic radiograph is recommended around age 6 or when the patient displays enough compliance for a diagnostic image. This allows for further assessment of the child's dental development as children diagnosed with SAS often have delayed eruption times, delayed development or missing multiple permanent teeth, and a significant delay in root development in permanent molars.
- The American Academy of Orthodontists (AAO) encourages an evaluation by an orthodontist of all children at no later than age 7. Because children with SAS sometimes experience a delay in the exfoliation of deciduous teeth, moderate-severe dental crowding, and lack of root development on permanent molars at a young age, we encourage families to have an evaluation consistent with the recommendations of the AAO. Care should be taken if performing maxillary expansion utilizing the 6-year molars, as root development can be minimal on these teeth in younger patients. There may be evidence to support the root development occurs into the late teenage years, but further research is needed to confirm.
- If accessible, a craniomaxillofacial team is beneficial for patients and families as it is typically composed of dental and medical specialists working together to provide the best possible care for patients. Coordination of care in an operating room setting is also highly beneficial as multiple services can participate in providing services for the child. A craniofacial team can be located by visiting https://acpa-cpf.org/team-care/team-listings/.

We are happy to provide further clarification and consult as needed regarding recommendations for a patient's plan of care.

Sincerely,

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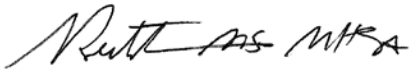

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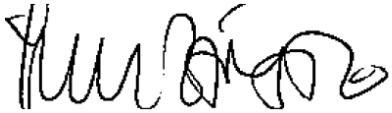

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