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Dental and Facial Characteristics of a Patient with Fetal Alcohol Syndrome – Case Report

Diagnostyka stomatologiczna pacjenta z alkoholowym zespołem płodowym FAS – opis przypadku

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A – research concept and design; **B** – collection and/or assembly of data; **C** – data analysis and interpretation; **D** – writing the article; **E** – critical revision of the article; **F** – final approval of article

Abstract

Fetal alcohol syndrome (FAS) represents a pattern of somatic as well as neurological congenital defects that can develop in children in cases of maternal consumption of alcohol during pregnancy. Fetal alcohol syndrome is an incurable disease, characterized in four categories: height, facial phenotype: moderate level of dysmorphic facial features characteristic of FAS, central nervous system (CNS) dysfunction features, probability of prenatal alcohol exposure. This paper describes the case of a 17-year-old patient with diagnosed strong myopathy with dysmorphic facial features (short palpebral fissures, wide-set eyes, low embedded, asymmetric ears, asymmetric and deformed ear auricles, a smooth philtrum and a thin upper lip), deficiency in body weight and height, microcephaly, abnormalities in sensorimotor progress and below-average intellectual development. The examination showed strong myopathy, horizontal and vertical shortening of the suprahyoid structures, a right-sided crossbite and impacted teeth. During the intraoral examination, it was impossible to establish a repetitive position of the mandible towards the jaw. The dental joint-muscle examination was performed by means of Bumann's manual functional analysis and posture analysis was also performed. Early diagnosis of FAS and proper dental diagnostics allow for effective and multi-targeted treatment of the patient **(Dent. Med. Probl. 2015, 52, 4, 505–511).**

Key words: alcohol, fetal alcohol syndrome, FAS, facial abnormalities, prenatal alcohol exposure.

Słowa kluczowe: alkohol, alkoholowy zespół płodowy, FAS, dysmorfia twarzy, ekspozycja alkoholu na płód.

Fetal alcohol syndrome is caused by maternal alcohol use during pregnancy and is one of the leading causes of preventable birth defects and developmental disabilities in both developed and developing countries [1, 2]. The first authors to describe FAS as a separate disease were Paul Lemoine et al. [3] in 1968, but the full spectrum of dysmorphic facial features was not described until 1973, when Jones and partners [3, 4] defined it in terms of "fetal alcohol syndrome". The irregularities resulting from ethanol use are generally called 'fetal alcohol spectrum disorders' (FASDs) [2]. For the majority of populations, FAS incidence affects 0.5–3/1000 live births [5], and the State Agency for Prevention of Alcohol-Related Problems (PARPA) estimates show that 3/1000 live births in Poland may be affected with FAS [6]. In high-risk groups (e.g., in surrogate families), FAS incidence is estimated to affect as many as 10–15/1000 live births [7]. In order to standardize guidelines for FAS diagnosis, in 2004 the University of Washington published a chart that—due to objective and quantitative measurement scales—made possible an increase in diagnosis precision and accuracy, thus ensuring better characteristics of the full spectrum of disorders, as well as quantitative measurement and a transparent recording system [8]. Table 1 shows a 4-Digit Diagnostic

Table 1. FAS diagnostics according to the 4-Digit Diagnostic Code

FAS diagnostics according to the 4-Digit Diagnostic Code			
1. Growth deficiency	2. Specific facial features	3. Brain functional abnormalities	4. Prenatal alcohol exposure
• body length at birth	• right and left palpebral fissure length	• structural abnormalities	• confirmed or unconfirmed
• body weight at birth	• distance between both inner eye corners	• neurological abnormalities	
• height and weight at particular ages	• presence of philtrum and upper lip stricture	• psychometric abnormalities	
		• interview with the child's caretaker (assessment of behavioral skills, ability to control emotions, sensorimotor integration, ability to think in the abstract, capacity to evaluate, memory, learning skills, information processing, spatial memory, social and adaptive skills, as well as motor control)	

Code, which reflects the magnitude of expression of the four key diagnostic features [2] (Table 1).

Each of the above-mentioned categories is ranked independently on a 4-point Likert scale, with 1 reflecting a complete absence of the FAS features and 4 reflecting a strong intensity of the classic patterns of the FAS features [2]. The above-mentioned dysmorphic features should be differentiated from other syndromes, e.g., Williams syndrome, maternal phenylketonuria, and chromosomal and deletion/duplication syndromes [9].

Case Report

A patient, 17 years and four months of age, with a diagnosis of fetal alcohol syndrome (FAS), presented to a dental office with dental issues. The documentation showed that the patient was born in the 35th week of pregnancy, with symptoms of deep birth asphyxia. Birth weight was noted to be 2100 grams and body length 49 cm, and there was no detailed data relating to prenatal and infantile development. Deficiency in body weight and height below the 3rd percentile was noted during early childhood (according to check-ups performed at the ages of two and four). At the age of four, the child's intelligence was examined against the Terman-Merrill test with a result of II = 69 (with II above 140 – very high intelligence, 139––120 – high intelligence, 119–110 – intelligence above average, 109–90 – normal or average intelligence, 89–80 – mental retardation, and 79–70 – borderline for mental impairment). The eight-year-old patient underwent orthodontic consultation and was diagnosed with right-sided crossbite and an oral pattern of breathing. The treatment was introduced with the use of removable braces, but the treatment was stopped after one month due to lack of cooperation on the child's end. At the age of 16, the patient's development was assessed by means of the 4-Digit Diagnostic Code chart (FASD) at the Diagnostic and Developmental Disorders Therapy Center.

The data gathered during the examination was put into four categories:

1. Height: 140 cm, below the 3rd percentile (clear result marker); weight: 36.1 kg, below the 3rd percentile (clear result marker); in general: considerable deficiency in height and weight.

2. Facial phenotype – moderate level of dysmorphic facial features characteristic of FAS: palpebral fissures – result significantly below – 2 SD (clear result marker); philtrum – ranked 4, which means – 1 SD (clear result marker); upper lip – ranked 3 (weak result marker).

3. Central nervous system (CNS) dysfunction features – probable CNS damage/dysfunction: head circumference – 49 cm (microcephaly), below the 3rd percentile (clear result marker). This was confirmed by the assessment of abnormalities in sensorimotor progress; below average intellectual development; retarded development of cognitive functions; recorded asymmetrical tonic neck reflex (ATNR), causing problems with making horizontal head movements independent from hand movements (difficulties in writing, reading and coordination); sensory disorders; hyperactivity; subsensitivity of vestibular system; problems with sensing distance and body coordination; auditory subsensitivity: and a distorted form of Moro reflex which may cause emotional difficulties (overly expressive reactions to stressful situations and sudden auditory stimulus) and hyperactivity.

4. Probability of prenatal alcohol exposure.

After the examination, the patient received

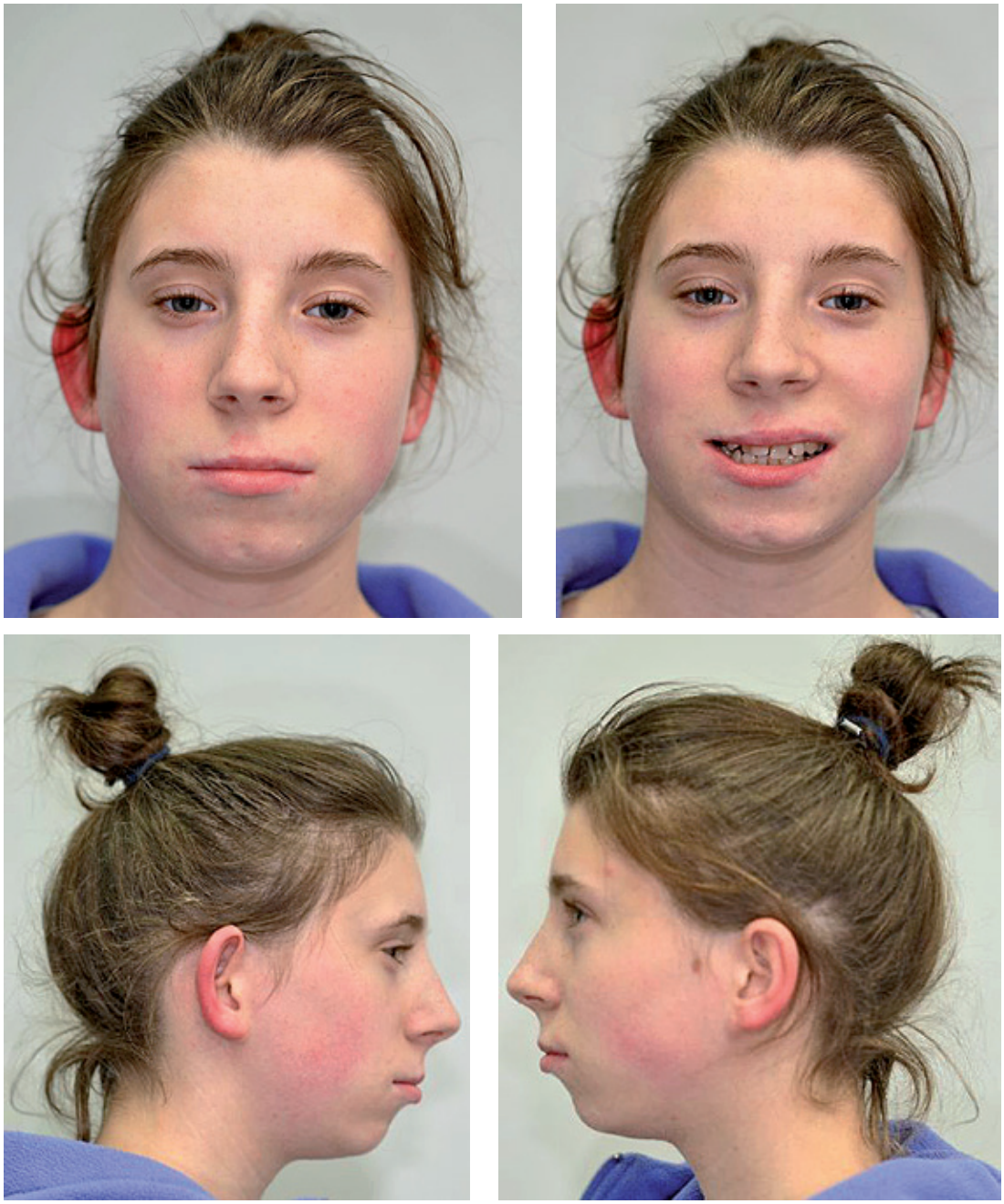

Fig. 1a–d. Extraoral examination photographs of FAS patient, taken at the age of 17 years and four months (a – en face, b – en face smiling, c – right profile, d – left profile). During the intraoral examination, it was impossible to establish a repetitive position of the mandible towards the jaw

the 4-Digit Code 4333 and, based purely on clinical features, was diagnosed with fetal alcohol syndrome, with probable alcohol exposure. During the dental check-up, the extraoral examination showed low embedded, asymmetric ears, asymmetric and deformed ear auricles, a short neck, wide-set eyes, drooping eyelids, short palpebral fissures, a snub nose, an increased distance between nose and mouth, a smooth philtrum and a thin upper lip (Fig. 1a–d).

During the intraoral examination, it was impossible to establish a repetitive position of the mandible towards the jaw.

Due to a suppurative fistula within the oral cavity vestibule, near to tooth 11 (a tooth injured during childhood years), the patient underwent a panoramic radiograph scan (Fig. 2) and endodontic therapy was planned. In addition, the patient received oral hygiene training, and due to the diagnosed oral pattern of breathing, and was ordered to perform orbicularis oris muscle exercises. People with FASD often have difficulty remembering regular routines, making plans and organizing themselves.

They may also have sensitivities to touch or to tastes or smells of products associated with

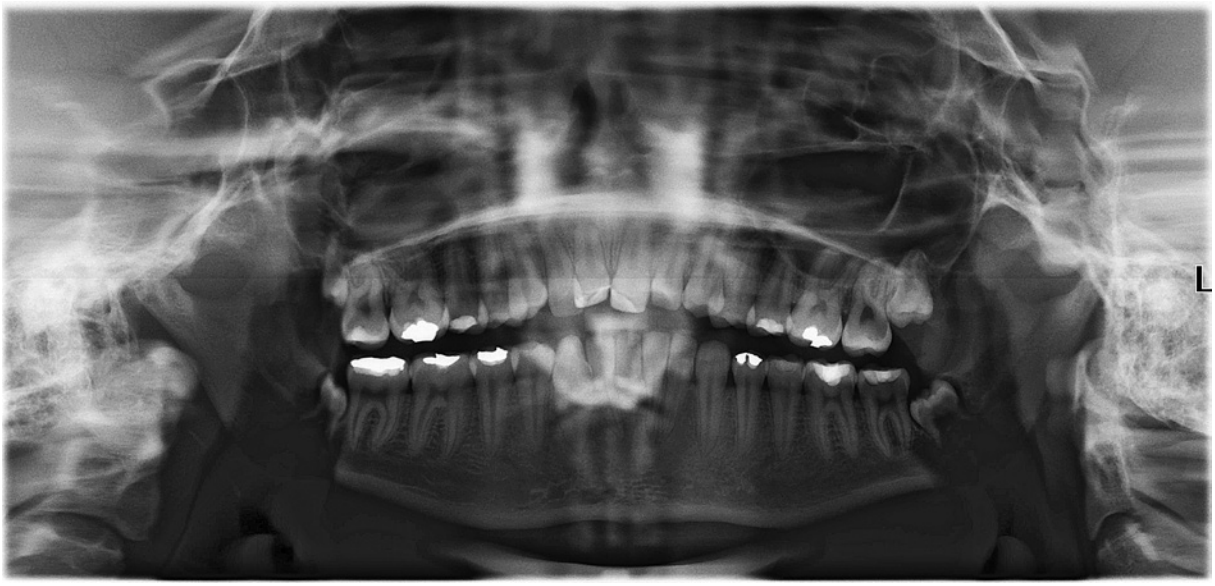

Fig. 2. Panoramic radiograph of patient with fetal alcohol syndrome (FAS), taken at the age of 17 years and 4 months

self care activities. There are some ways that may help support self care routines. Encourage the use of toothpaste, floss and be specific about how often and long to brush. Poor dental hygiene is often a significant health issue which is heightened because toothbrushing requires a great deal of fine motor coordination and involves sensory issues.

Due to significant speech disorders, referral to a speech therapist was issued, and the surgical removal of four retained third molars was recommended. The speech therapist diagnosed spontaneous speech, a fast speech rate and occlusal seizure, especially in the medial part of words. Furthermore, the letter "r" in articulation sounded uvular or vibrational, and there was a slight nasal quality to oral sounds.

The patient, due to her frequent headaches, was referred for neurological consultation and a joint-muscle examination of the viscerocranium. The dental joint-muscle examination was performed when the patient was 17 years and eight months old, by means of Bumann's manual functional analysis (10), and posture analysis was also performed.

The manual functional analysis (MFA) described by Bumann allows for a non-invasive and accurate diagnosis of functional disorders of the masticatory apparatus. This analysis comprises anamnesis, preceded by a patient questionnaire (including questions regarding possible symptoms of functional disorders of the masticatory apparatus) and a physical examination. In the physical examination, the path and range of mandibular movements are investigated (abduction and adduction, protrusive and lateral movements). The condition of the articular surface of the upper and lower parts of the temporomandibular joint is then examined, and following this, the articular capsule and ligaments are checked. An isometric test and palpation are then used in order to assess the mandibular muscles and the length of the suprahyoid structures. The MFA also makes it possible to assess clicking and popping of the temporomandibular joint and establish reasons for their occurrence.

The medical interview showed a parafunction in the form of gum chewing two hours daily, and the occurrence of frontal lobe headaches several times a week. The examination showed strong myopathy, horizontal and vertical shortening of the suprahyoid structures and postural deformity. Due to the myopathy, it was impossible to define the neuromuscular location of the mandible. After each adduction, the patient put her mandible in a different position, thus making it impossible to define the type of occlusal problems and a proper orthodontic diagnosis. It was also impossible to establish a repetitive relation of the mandible towards the jaw and a centric relation of the condyle in the temporomandibular joint. The muscle palpation showed the presence of 26 points of maximum painfulness (7–10 on the 0–10 scale), with as many as 22 points reaching level 10 (on the scale 0–10, where 0 means lack of pain, and 10 means unbearable pain). Level 10 at just one of the points constitutes an absolute indication for occlusal splint therapy. Based on the established functional diagnosis, a treatment was introduced that consisted of making a Michigan-type occlusal splint, founded on the lower dental arch, and the simultaneous referral for physical therapy. As part of the physical therapy, the pa-

tient was ordered to perform exercises to correct the asymmetry of back muscle tension and the excessive contracture of pelvic muscles. As part of the mandibular muscle physiotherapy, the patient performed 30 repetitions of Gerry exercises three times a day, with her tongue placed on her palate behind the central incisors. The treatment using the Michigan-type occlusal splint lasted for six months and consisted of the correction of the splint leading plane after the first and fifth weeks of splint usage. The patient postponed consecutive control visits and, as a consequence, the next correction of the splint leading plane was performed 18 weeks after the last control. A significant decrease in the occurrence of headaches was noted as a result of the associated occlusal splint therapy and physiotherapy. After just five weeks of treatment, the abduction and adduction path of the mandible became repetitive. A comparative clinical study was performed by means of manual functional analysis, after six months of occlusal splint therapy and physiotherapy, which showed an increased mandible movement range, significant improvement in muscle condition, and a decreased occurrence of tension-type headaches. The muscle palpation showed 0 points of maximum painfulness. The isometric test performed on the mandibular muscles showed no occurrence of muscle fatigue symptoms. A visible improvement in the condition of the suprahyoid structures was also noted (horizontal and vertical shortening of these structures had diminished). The associated treatment consisting of the use of a Michigan-type occlusal splint and physiotherapy made it possible to determine a repetitive mandible position towards the jaw, as well as to find and register a centric relation of the condyle in the temporomandibular joint (according to Okeson's definition of centric relation) (11), thus allowing the determination of a proper starting point for the planned orthodontic therapy. The intraoral examination confirmed the previous diagnosis regarding a right-sided crossbite and impacted teeth in the upper and lower dental arch (Fig. 3a–e).

A further treatment plan includes orthodontic treatment for malocclusion. As a consequence of the neurological consultation, the patient was referred for MRI, which confirmed the microcephaly and altered relation of the neurocranium and viscerocranium sizes, as well as hypoplasia of the cerebellar vermis. The patient is under constant specialist care in the field of psychiatry, neurology, endocrinology, speech therapy and orthopedics, due to the diagnosed scoliosis. Currently the patient does not receive any medications.

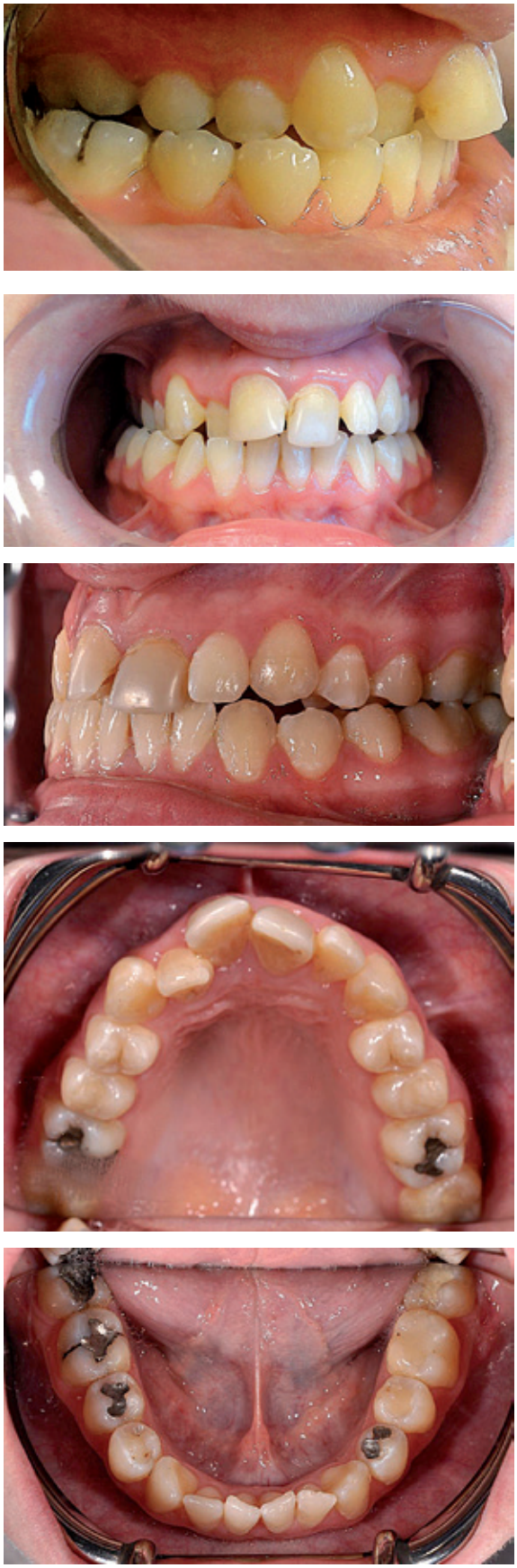

Fig. 3a–e. Intraoral photographs of patient with fetal alcohol syndrome (FAS), taken at the age of 17 years and four months (a – right side, b – en face, c – left side, d – occlusal projection of upper dental arch, e – occlusal projection of lower dental arch)

Discussion

Nowadays, the basic difficulties in diagnosing patients with fetal alcohol syndrome include the lack of clinical standardization in defining FAS and other disorders caused by prenatal alcohol exposure, as well as the lack of objective and quantitative measurement scales and intensity scales to record key symptoms characteristic of FAS [8]. As a consequence, descriptions of the spectrum of disorders relating to FAS have been significantly different between particular researchers [1, 5, 12–14]. From a clinical standpoint, an erroneous diagnostic classification can lead to incorrect patient care, an increased risk of secondary disorders [15], and incorrect estimates of data regarding the frequency of FAS occurrence [5]. There are no two medical cases diagnosed with fetal alcohol syndrome that show the same combination of disabilities and other irregularities; that is why, in the case of this patient, the 4-Digit Code FAS diagnostic chart was extremely helpful, as it increased diagnosis precision and accuracy. Due to the dysmorphic facial features, it is easiest to diagnose fetal alcohol syndrome in patients between eight months and 10 years of age. In newborns, the characteristic features are not yet fully developed, and after the tenth year of life until adulthood, they gradually diminish. In the case under discussion, the patient was 17 years old and despite being diagnosed with full-blown FAS, certain dysmorphic facial features were not being noted. During a child's school years, certain emotional disorders make it easier to diagnose FAS, and abnormalities in head morphology are noticeable at a later stage [16]. The introduction of the associated treatment, consisting in the use of a Michigan-type occlusal splint and physiotherapy, made it possible to find and register a repetitive mandible position towards the jaw and to diagnose malocclusion in the form of a right-sided crossbite, just as in the case of the patient described by Szymoniak and Jankowska-Wika [17]. These two cases also shared similarities between the escalation of behavioral changes in the form of mental impairment, speech disorders and hyperactivity. The diagnosis of FAS is a challenge, and in order to precisely interpret the wide range of associated features and make an accurate diagnosis, it requires a multidisciplinary approach, including the involvement of a pediatric dentist, a psychologist, and speech and occupational therapists [8].

By increasing the knowledge available concerning the wide spectrum of clinical disorders, both dental and general, that characterize fetal alcohol syndrome, it is easier to efficiently diagnose and treat FAS, regardless of the patient's actual age.

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