Autism Disorder (AD): An Updated Review for Paediatric Dentists

INTRODUCTION
Autism disorder (AD) was first described in 1943 by the American child psychologist, Leo Kanner. He presented 11 children whose behaviours were obviously different from those of others. Kanner suspected that they had an inborn feature which had prevented their regular social contacts [1]. Autism Disorder is sometimes referred to as early infantile autism, childhood autism, or Kanner’s autism.

Autistic Disorder is categorized in the DSM-IV (Diagnostic and Statistical Manual of Mental Disorders, 4th ed) under the section, Pervasive Developmental Disorders (PDD), which is also referred to as Autism Spectrum Disorder (ASD). It is characterized by abnormal emotional, social behaviour and linguistic development [2]. The term “PDD” is widely used by professionals to refer children with autism and related disorders; however, there is a great deal of disagreement and confusion among professionals concerning the PDD label. Diagnosis of PDD, including autism or any other developmental disability, is based upon the Diagnostic and Statistical Manual of Mental Disorders – Fourth Edition (DSM-IV), published by the American Psychiatric Association (Washington, DC, 1994), and it is the main diagnostic reference of mental health professionals in the U.S. [3,4]. According to the DSM-IV, the term, “PDD” is not a specific diagnosis, but it is an umbrella term under which the specific diagnoses are defined, which include:

**Autistic Disorder (299.00 DSM-IV)**
People with autistic disorder (or “classic” autism), the most severe form of ASD, have significant difficulty in talking and relating to others, they display compulsive and repetitive behaviours, and they have intellectual disability [5].

**Asperger Syndrome (299.00 DSM-IV)**
People with Asperger syndrome display milder symptoms of autistic disorder. They may have social challenges and unusual behaviours, but they have no significant problems with language or intellectual disability [5].

**Pervasive Developmental Disorder Not Otherwise Specified (PDD-NOS; or “atypical autism”) (299.80 DSM-IV)**
People who display some classic symptoms of autistic disorder or Asperger syndrome but do not meet the criteria for a specific diagnosis, may be diagnosed with PDD-NOS. Characteristics of PDD-NOS include impaired communications and social interactions in varying degrees, sensitivities to sights and sounds, and repetitive and stereotyped patterns of behaviour [5].

**Rare Disorders: Rett Syndrome and Childhood Disintegrative Disorder (CDD) (299.80 DSM-IV)**
Rett syndrome occurs almost exclusively in females who are between 6 and 18 months of age. Characteristics include poor head growth, regression of mental and social development, and avoidance of social contact and excessive hand and foot activity. CDD develops in children who initially seem normal for the first two years and then suddenly exhibit a disintegration of language, motor, and social skills; fail to make friends; lose bowel and bladder control; and develop seizures [5].

Being the broadest and most studied group in this spectrum, the Autism Disorder (AD) has been reviewed in this article. It is a lifelong neurodevelopment disorder and has no cure [6]. It is highly heritable, although the genetics of autism is complex.

**Definition**
Autism Society of America (ASA) [7] defined AD as: A complex developmental disability that typically appears during the first 3 years of life and is the result of a neurological disorder that affects the normal functioning of the brain, impacting development in the areas of social interaction and communication skills.

The National Institute of Child Health and Human Development defined Autism Disorder (AD) as:
“A complex biological disorder that generally lasts throughout a person’s life, as it starts before age three, in the developmental period, and causes delay or problems in many different ways in which a person develops or grows.”

**Aetiology**
Considering the complex nature of autism, a single cause is unlikely. Possible aetiologies of autism are

**Genetics**
Gene mutations, gene deletions, copy number variants (CNVs) and other genetic anomalies are all persuasively linked to autism [8].
Autism affects approximately three times as many males as females. Taken together with studies which have looked at families and twins, the results suggest that autism has a genetic component [9]. It is likely that a number of genes are involved. However, the exact mechanism by which genes are implicated in autism is unclear and this is an important focus for future research. In addition, future studies aim to determine how genes interact with environmental factors in autism [6].

**Biology of Brain**

Strong evidence suggests that AD is an organically based neurodevelopmental disorder which is associated with abnormalities in brain structure and function. Characteristic findings are a

i. Reduced number of purkinje cells in the posterior inferior regions of the cerebellar hemisphere.

ii. Truncation in the dendritic tree, development of neurons in the limbic system [8,10].


iv. Size of the brain stem structures and the entire cerebellar vermis and their components were significantly smaller in an autistic group than in control group, which was found on using magnetic resonance imaging [12].

**Prenatal Factors**

Intrauterine viral infections or metabolic disorders may play an important role in the pathogenesis of AD [12]. Intrauterine exposures to the teratogenic drugs, thalidomide and valproate have been implicated as the cause of autism in few affected children [13].

**Coexisting Medical Conditions**

i. Seizure disorder

ii. Fragile X syndrome is found in 2-5 % of autistic individuals and it represents the largest known subgroup of patients with AD, with known aetiology [14].

iii. Tuberous sclerosis complex (TSC): 0.4-3% of patients with AD had TSC and 17-58 % of subjects with TSC had AD [15].

iv. Autism patients have allergies, immune system problems, gastrointestinal disturbances and seizures. Dentists must be aware of these co-morbid conditions so that they can give optimal care to the children with autism disorder [16].

v. Neuropsychiatric disorder: Developmental delay, dysmorphic features, obstetric complications, an unequal sex ratio, and extremes of head size represent nonspecific signs of autism [17].

**Prevalence Rate of Autism**

Autism is a global health crisis that knows no borders - it does not discriminate individuals, based on nationality, ethnicity or social status. Centers for Disease Control and Prevention, or CDC, conducts year-round studies that relate to and indicate the incidence and prevalence of autism. They also provide systematic public health monitoring of a wide range of diagnosed disorders. Their research is held to the highest standards of validity and reliability. The CDC defines persons with Autism Spectrum Disorder (ASD) as those who demonstrate “impairments in social, communicative and behaviour development, that typically are present before age of 3 years and are often are accompanied by abnormalities in cognitive functioning, learning, attention and sensory processing” (Centers for Disease Control and Prevention, 2011) [18].

Worldwide, population-based studies which were conducted before 1985 have identified the prevalence of autism and related conditions among children who were under 18-years-old, to be approximately 0.5 per 1,000 children. The CDC’s most recent studies suggest that children who meet the “Autistic Disorder” criteria range in numbers of up to 12 per 1,000 children, worldwide. In the USA, rates of autism climbed “greater than tenfold” from rates which were identified during the 1980s, to a current estimate of 1 in 110 children (1%) [19].

Prevalence rates per country are difficult to determine. Posserud et al., [20] and Wong [21] calculated and identified respective country’s approximate prevalence rates of autism diagnoses of children living in other countries throughout the world and combined as follows:

- Australia: 6.25 in 1000
- China: 1.1 in 1000
- Denmark: Nearly 9 in 1000
- India: 1 in 250
- Japan: Nearly 3 in 1000
- Mexico: 2 to 6 per 1000
- Canada: 1 in 154
- Sweden: 1 in 188
- Finland: 1 in 833
- Denmark: 1 in 833
- Iceland: 1 in 769
- Philippines: 500,000 children, total

Given this statistic, it is likely that dentists will treat patients with an AD in their practice. Therefore, having an understanding of AD is essential for planning effective patient treatment.

**Diagnosis of AD**

ADs are diagnosed by looking at a child’s behaviour and development. No medical tests such as blood tests or biopsies can be performed to diagnose AD. Diagnosing AD involves a two step process; first step is a developmental screening, followed by a comprehensive diagnostic evaluation. A general developmental screening is performed for children during regular paediatric check-ups. The screening determines whether children are learning basic skills, such as learning skills, speech, behaviours, and movement, at the appropriate time. Age intervals for developmental screenings are 9, 18, and 24 to 30 months. Developmental screenings which are specifically designed to look for AD should be performed at 18 and 24 months of age. Examples of such screenings are the Modified Checklist for Autism in Toddlers (M-CHAT) and Screening Tool for Autism in Toddlers and Young Children (STAT).

After a developmental screening, the next step for diagnosing AD is a comprehensive diagnostic evaluation. This incorporates a thorough review of the child’s behaviour and development, interview with the parents, hearing and vision tests, and genetic and neurological tests. The evaluation may require referrals to specialists such as developmental paediatricians, child neurologists, psychologists, and psychiatrists. Comprehensive testing will help in ruling out other conditions or disorders such as hearing loss which is unrelated to AD [22].

The National Autism Plan for Children (NAPC) [16] has provided a template for professionals. The diagnostic criteria for autism [Table/Fig-1] require the presence of 6 symptoms from 3 categories:

- Impaired reciprocal social interaction (at least 2 symptoms),
- Impaired communication (at least 1 symptom),
- Restricted, repetitive, or stereotyped behaviours (at least 1 symptom).

These criteria reflect the central role of deficits in social behaviour in children with ADs.

Early diagnosis of AD has many advantages for the child and family. These include [3]:

- Early information about education and support;
- Earlier access to targeted social, communication, behavioural interventions
A. A total of 6 items from the following criteria 1, 2, and 3, with at least 2 from criterion 1 and each from criterion 2 and 3
1. Qualitative impairment in social interaction as manifested by at least 2 of the following:
   a. Marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body posture, and gestures to regulate social interaction
   b. Failure to develop peer relationships appropriate to developmental level
   c. Lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g. lack of showing, bringing, or pointing out objects of interest)
   d. Lack of social or emotional reciprocity
2. Qualitative impairments in communication as manifested by at least 1 of the following:
   a. Delay in or total lack of development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)
   b. In individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation.
   c. Stereotyped and repetitive use of language or idiosyncratic language
   d. Lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level.
3. Restricted, repetitive, and stereotyped patterns of behavior, interests, and activities as manifested by at least 1 of the following:
   a. Encompassing preoccupation with 1 or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
   b. Apparently inflexible adherence to specific, nonfunctional routines or rituals
   c. Stereotyped and repetitive motor mannerisms (e.g. hand or finger flapping or twisting, or complex whole-body movements)
   d. Persistent preoccupation with parts of objects
B. Delay or abnormal functioning in at least 1 of the following areas, with onset before age 3 years:
1. Social interaction
2. Language as used in social communication
3. Symbolic or imaginative play
C. Disturbance not better accounted for by Rett’s disorder or childhood disintegrative disorder

Identification of co-morbid medical, developmental and psychiatric conditions.

Clinical Presentation
The main challenge to the dental team may be the reduced ability of autistic patients in communicating and relating with others; further problems include
1. Uneven intellectual development.
2. Peculiar repetitive body movements.
3. Hyperactivity.
4. Limited attention span.
5. Low frustration threshold that may lead to temper tantrums or bizarre vocalization [23].
6. Although there appears to be no experimental verification, several publications have described autistic individuals as having high threshold to pain, which have concluded that short procedures may be carried out without local anaesthesia [24].
7. AD exhibits tactile and auditory hypersensitivity
8. It also exhibits exaggerated reactions to light and odours, so team should be prepared for unpredictable and unusual responses to sensory stimuli [3].
9. Some authors attribute a strong urge for soft, sticky, and sweet foods.
10. Patients with AD tend to dislike changes in their environment and need sameness and continuity [25].
11. They may react with tantrums over small environmental changes [26].
12. Self-injurious behaviour (SIB)
SIB occurs in 4-5% on individuals with different psychiatric conditions, especially in those with AD and brain damage. A change in daily routine may initiate or increase it. SIB may range from self-pincching or scratching to severe self-biting or head banging.

An injury might be done to either attract the attention of a family member or clinician or to avoid unwanted events. The prevalence of these lesions was 13% [27] but it was far lower than that seen in other studies, in the order of 68% [28].

Oral Health Status
Patients with AD do not present very specific oral disorders. They pose the greatest challenge for dentists, due to their complex and varied clinical manifestations.

Dental Caries
Shapira et al., [29] evaluated DMFT and found that institutionalized autistic individuals exhibited lower caries rate than institutionalized schizophrenics. The caries rate of autistic children in a day care facility was similar to that of their peers.

Low and Lindemann [30] assessed dental needs of AD patients by studying a group of 20 AD subjects and compared them to 20 nonautistic, age-matched controls. In the primary dentition, the patients with AD demonstrated a significantly higher caries rate (dmf) than the controls during initial examination, but at recall examinations, dmf values were comparable. In patients with permanent dentition, both at baseline and recall, Dmf scores were not different between the groups. A low incidence of dental caries was mentioned by Kamen and Skier [31].

Kopel stated that patients with AD did not exhibit any unique features of hard or soft intra- or periradial tissues and that the prevalence of dental disease in them was similar to that seen in other children [32]. In a recent study, autistic individuals were compared with non-autistic healthy controls and they were found to have neither a higher salivary flow rate nor a better buffer capacity of the saliva and similar dental caries experiences were observed in both primary and permanent dentitions [33].

In general, children with autism prefer soft and sweetened foods and they tend to pout food inside the mouth instead of swallowing it, due to poor tongue coordination, thereby increasing the susceptibility to caries [14]. Moreover, the risk for dental caries can be expected to be higher in these patients due to difficulties in brushing and flossing, though the oral hygiene of these subjects is known to be deficient [34]. Nevertheless, many authors have found that the prevalence of caries had no difference on comparing autistic and non-autistic individuals [35], and in some cases, the prevalence of caries in children with AD could even be comparatively lower [29,31,36].

Periodontal Status
Majority of autistic children had poor oral hygiene, and almost all of them had gingivitis [35]. These changes could be related to irregular brushing habits, because of the difficulties the trainers and the parents encountered when they brushed the children’s teeth. They could also be caused by lack of the necessary manual dexterity of autistic children, which may have resulted in inadequate tooth brushing. Furthermore, the findings of this study reflected poor dental awareness, a lack of dental education and deficiency in receiving oral hygiene instructions from dental staff. Another possible explanation for the presence of generalized gingivitis might be the side effects of medications which were used to control the manifestations of autism, such as psychoactive drugs or anticonvulsants, with the most common drug classes being antidepressants, stimulants, and antipsychotics [32].

Oral Habits
Harmful oral habits are common, which consist of bruxism, tongue thrusting, picking at the gingiva and lip biting. Bruxism or forceful grinding of teeth is one of the sleep problems which are commonly observed in children with autism. Dentist can recommend a mouth guard to stop this self-injurious behaviour. Even though the
communications and behavioural problems in children with autism pose challenges for the dentist, treatment given with proper planning and a lot of patience can definitely make a difference [37].

Tooth Eruption
Tooth eruption may be delayed due to phenytoin-induced gingival hyperplasia. Phenytoin is commonly prescribed for people with autism [38].

Dental Injury
The rate of dental injuries is higher among autistic children. The most common dental injury was enamel fracture and the most frequently injured teeth were the permanent maxillary central incisors [39].

Malocclusion
Patients with ASD do not present very specific malocclusions, still they do show a greater tendency towards certain malocclusions (e.g., ogival palate and anterior open bite [40].

Dental Management Considerations
AD is a heterogeneous disorder with a wide range of expressions. Therefore, treatment approaches that may yield a positive outcome in one patient may prove to be ineffective for another. Also, there is a limited number of studies that have addressed basic behaviour management techniques and procedural modifications with regards to dental treatment of children with AD.

Appointment Structure
Because of the limited attention span of AD patients short, well-organized appointments should be planned and the waiting time should not exceed 10-15 minutes, to avoid upsets [41].

Dental Environment
Discussion of any aspect of the actual work should be avoided during the course. Light background music might be beneficial. Anyone participating in the procedure should minimize movements, because an autistic child can be easily distracted [42].

Applied Behaviour Analysis (ABA)
Applied behaviour analysis is a branch of psychology that is focused on the analysis and modification of human behaviour. ABA practices are based on the basic principles which were developed by Skinner. Baer and colleagues provided an in-depth explanation of the characteristics of ABA. Procedures based on ABA are evidence based and they have been accepted by the American Academy of Paediatrics in the management of AD. In dentistry, the use of these procedures has the potential to improve the results of traditional behaviour management procedures [43]. By increasing the likelihood of patients who accept simple and routine dental procedures; dentists can decrease the need for more intrusive procedures such as restraints and sedation [44].

Visual Pedagogy
Bäckman and Pilebro [45] in a prospective study which included clinical examinations and structured interviews, produced a series of pictures that showed a structured method and technique of tooth brushing. The pictures were placed in the bathroom or wherever tooth brushing was performed. Fourteen children with autism, who were aged between 5 and 13 years were involved. After 12 months, the amount of visible plaque was reduced. After 18 months, most of the parents found maintaining good oral hygiene easier than they had found it before the study and concluded that visual pedagogy was a useful tool for helping people with autism in improving their oral hygiene.

Communicative Behaviour Management Techniques
Techniques which are commonly advocated and used [30-32] for behaviour modifications in patients with AD are the same as those which are used for nonautistic individuals: tell, show, do and immediate, frequent positive and negative reinforcement, paired with firmness, wherever necessary. However, higher rate of flexibility is required to comply with quickly changing patient needs. Other recommendations which are again based on the modeling effects of constant positive reinforcers, are immediate verbal praise after each accomplished step of a procedure and a prize at the end of a dental session. The oral communication should be carried out in clear, short, and simple sentences [46]. Inappropriate behaviour should be ignored [47] Hand over mouth was not considered.

Pharmacological Behaviour Management Techniques
Several authors [48] have described the use of pharmacological agents. Frequently used drugs were nitrous oxide, diazepam, hydroxyzine, chloral hydrate, and promethazine, in contrast to chlorpromazine, diphenhydramine, and meperidine. The drugs were administered in different dosages and regimens, as a sole agent or in various combinations. In some patients, several different regimens and combinations were attempted, in order to be successful. Reported success rate varied from a limited 30 to 70% [49,50]. A lengthier administration and higher concentrations of nitrous oxide than usual were required to achieve the desired level of sedation in patients with AD. Giving treatment in the operating room by using general anaesthesia was considered only if all other approaches had failed.

LEGAL CONSIDERATIONS
Autism is a disability under the Disability Discrimination Act. According to this legislation, organizations must make reasonable adjustments to include autistic individuals; under the Disability Equality Duty, all public bodies must promote the equality of autistic individuals. As autism affects social communications and interactions, social inclusions, for instance, in leisure activities, are a particular challenge.

The range of ability and disability in autism means that individuals and their families have diverse needs. Their primary needs will change with age from early childhood and educational services, to support in adulthood, for example with employment.

SUMMARY
1. There is strong evidence that AD is caused by a pre or perinatal insult and that it is not a progressive degenerative process. Although males are almost four times more affected, no moderate-to-strong gene effect on the X chromosome has been detected.
2. Early (prior to 40 months of age) detection is important, because early therapy results in faster and greater improvement than later intervention. Paediatric dentists are well-suited to be primary health care providers for early screening of the risk group, because paediatric dental care ideally starts by age 1 and because they are particularly trained in the treatment of individuals with special needs.
3. Autistic individuals generally do not display specific dental findings, but compromised oral hygiene can contribute to an increased risk for caries and especially periodontitis in some patients; patients taking phenytoin or phenobarbital for seizure control are at a risk for gingival overgrowth.
4. Tell-show-do, voice control with short, clear commands and positive reinforcement are successful first-line management techniques for autistic patients.
Thus, to conclude, an autistic patient can often present a considerable challenge to the dentists and staff. Patience and time are vital while working with these children.

'The test of mortality of a society is what it does for its children’

Dietrich Bonhoeffer

REFERENCES


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