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Review Article

Oral Health Status In Intellectually Disabled - A Review

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ABSTRACT

Mental health envisages one's ability to realize one's intellectual and emotional potential. Intellectual disability refers to a group of disorders characterized by significant limitations both in intellectual functioning and adaptive behavior. The oral health of people with learning disabilities is often compromised, which has a deleterious effect on their well-being and quality of life. Dental caries and periodontal disease are among the most common secondary conditions affecting people with intellectual disabilities (ID). This literature review outlines oral health status among intellectually disabled [Mentally retarded (MR) and Down Syndrome (DS)]. The individuals with MR have poorer overall oral health and oral hygiene compared with the general population. Though the prevalence of dental caries is almost similar to general population the periodontal conditions are poor. Numerous barriers have been identified which could have profound effect on oral health of these individuals. Individuals with Down syndrome have less carious teeth, but experience more severe and extensive periodontal diseases. They have more missing, misaligned teeth and often affected with malocclusion. In conclusion, individuals with ID have poorer oral hygiene and higher prevalence with greater severity of periodontal disease. Dental caries was similar or low than general population but the prevalence of untreated caries was high. It is important for dental professionals to identify and quantify their needs and plan oral health promotion programmers for better oral function and an improved quality of life.

Key words: caries experience, dental caries, Down's syndrome, intellectual disability, oral health status, mental retardation, periodontal disease.

INTRODUCTION

Health of an individual and health of a society are recognized as being interrelated. [1] Physical, mental and social health is vital and interwoven strands of life. [2] Mental health is more than the mere lack of mental disorders. Concepts of mental health include subjective well-being, perceived self-efficacy, autonomy, competence, intergenerational dependence and recognition of the ability to realize one's intellectual and emotional potential mental health problems affect society as a whole, and not just a small, isolated segment. [2]

Oral health is an integral element of general health and well-being. Good oral health enables individuals to communicate effectively, to eat and enjoy a variety of foods, and is important in overall quality of life, self-esteem and social confidence. [3] The oral health of the disabled may be neglected because of the mental disability, a demanding disease or limited access to oral health care. Moreover, because of their level of function and their limited ability to undergo an oral examination, the mentally disabled present specific challenges when their oral health is assessed. [4] This overview outlines, epidemiology, and

classification, clinical features, along with management of individuals suffering from Intellectual Developmental disorders.

Intellectual Developmental Disorders

Many terms and definitions are used to refer to intellectual developmental disabilities, such as mental retardation, mental handicap, intellectual disabilities, and learning disabilities. [5] Nevertheless, all these definitions have three criteria in common: significant limitations in intellectual functioning, significant limitations in adaptive behaviour, and manifestation of these symptoms before adulthood. [5]

According to Medical Subject Heading (MeSH) intellectual disabilities are classified under neurobehavioral manifestations. Intellectual Disability includes Cri-du-Chat Syndrome, De Lange Syndrome, Down syndrome, Mental Retardation(X linked), Prader-Willi Syndrome, Rubinstein- Taybi Syndrome, WAGR Syndrome, Williams Syndrome. Pubmed search reveals that most of the studies in this area are done among subjects with Mental retardation and Down syndrome which are dealt in detail in this review. Whereas limited or no studies regarding oral health and Cri-du-Chat Syndrome, De Lange Syndrome, Prader-Willi Syndrome, Rubinstein- Taybi Syndrome, WAGR Syndrome, Williams Syndrome is available. Hence literature pertaining to Mental Retardation and Down's syndrome are included in this review.

Mental Retardation (MR)

Internationally the definition of Mental Retardation has moved away from medical model to rehabilitative model. Current trend is to describe the condition by using functional and educational terms rather than clinical terms. Definitions are listed chronologically to demonstrate the variations in describing condition of Mental Retardation. [6]

As per American Association on Mental Deficiency (AAMD), 1983 Mental retardation refers to "a significantly sub-average general intellectual functioning resulting in or associated with concurrent impairments in adaptive behavior and manifested during the developmental period". [6]

It is a more functional definition which stresses the interaction between the person's capabilities, the environment in which the individual functions, and the need for support systems. [6]

American Association of Mental Retardation (AAMR) in 1992, has defined MR as the onset of significant limitations in both general intellectual and adaptive functioning during the developmental period (18 years and under). This refers to significantly sub-average intellectual functioning, existing concurrently with or more of the following applicable adaptive skill areas, communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academics and leisure. [6]

American Psychological Association (APA) in 1994 in the Diagnostic and Statistical Manual of Mental Disorders (DSM) -IV gave three diagnostic criteria for MR, including sub-average intellectual functioning (IQ < 70), impairments in adaptive functioning and onset before age 18. [7]

AAMR in 2002 redefined MR as "A disability characterized by significant limitations, both in intellectual functioning and in adaptive behavior, as expressed in conceptual, social and practical adaptive skills, the disability originating before the age of 18 years". [6]

In the year 2009 AAMR was rechristened as **American Association on Intellectual and Developmental Disabilities (AAIDD).** Hence defined **Intellectual disability** as "a disability characterized by significant limitations both in intellectual functioning and in adaptive behavior, which covers many everyday social and practical skills. This disability originates before the age of 18 years." [6]

Intellectual disability involves impairments of general mental abilities that

impact adaptive functioning in three domains, or areas. These domains determine how well an individual copes with everyday tasks:

The **conceptual domain** includes skills in language, reading, writing, math, reasoning, knowledge, and memory.

The **social domain** refers to empathy, social judgment, interpersonal communication skills, the ability to make and retain friendships, and similar capacities.

The **practical domain** centers on self-management in areas such as personal care, job responsibilities, money management, recreation, and organizing school and work tasks. [6]

Epidemiology [2, 8-10]

→ About 3% of the world population is estimated to be mentally retarded-1.5 times more common among men than among women. Prevalence of severe mental retardation is about 3 per 1.000 population and 30 per 1.000 for mild mental retardation. High mortality among subjects with severe & profound mental retardation is due to associated physical disease.

It is more common in developing countries because of the higher incidence of injuries and anoxia around birth, and early childhood brain infections. According to the WHO, the true prevalence rate of total MR in industrialized countries comes close to 3%"; in the United States rate is 1%-3%, whereas the Scandinavian countries claim that the 1 % figure is their true prevalence. In India, Mentally retarded population accounts for 0.44 million individuals and 11.34% of total disabled**.**

Etiology [7]

1. Chromosomal abnormalities

Down's syndromes, Fragile X syndrome, Klinefelter's syndrome (47, XXY), Turner's syndrome, Cat-cry syndrome, Prader-Willi syndrome and de Lange's syndrome

2. Prenatal problems

Cytomegalovirus infection, Toxoplasmosis, Herpes, Syphilis, Rubella, Human, Immunodeficiency Virus, prolonged maternal fever in the first trimester, exposure to anticonvulsants or alcohol and untreated maternal phenylketonuria.

3. Perinatal problems

Late pregnancy complications, diseases in mother such as heart and kidney disease and diabetes and placental dysfunction, during delivery (labour) severe prematurity, very low birth weight, birth asphyxia, difficult and/or complicated delivery and birth trauma), neonatal (first 4 weeks of life) septicemia, severe jaundice, hypoglycemia.

4. Postnatal problems (in infancy and childhood)

Brain infections such as tuberculosis, Japanese encephalitis, and bacterial meningitis. As well as head injury, chronic lead exposure, severe and prolonged malnutrition and gross under stimulation.

5. Metabolic disorders

Phenylketonuria, hypothyroidism, mucopolysaccharidosis, sphingolipidoses

6. Exposure to certain types of disease or toxins

Exposure to poisons like lead or mercury

7. Iodine deficiency (cretinism)

8. Malnutrition

Classification [11]

The term "mental retardation" as mentioned in ICD-10 and DSM-IV classification systems is now referred as to intellectual developmental disabilities as per DSM-V classification systems.

In addition, the parenthetical name (intellectual developmental disorder) is included in the text to reflect deficits in cognitive capacity beginning in the developmental period. Together, these revisions bring DSM into alignment with terminology used by the World Health Organization's International Classification of Diseases, other professional disciplines and organizations, such as the American Association on Intellectual and Developmental Disabilities, and the U.S. Department of Education.

Based on the 1983 AAMR definition, the operational classification for persons with mental retardation is as follows **:(Table-1)** [7,12 -14)]

Table 1: IQ ranges for different levels of mental retardation

Level of Retardation	IQ range	
	Stanford-Binet and Cattell Tests	Wechsler Scales
Mild	52-67	55-69
Moderate	36-51	40-54
Severe	20-35	25-39
Profound	0-19	0-24

According to ICD-10 guide for mental retardation, based on Severity of Retardation and Problem Behaviours, axis I is used to record codes from Section F7 of the classification. Severity of retardation is recorded with a second digit code (F70 to F79). [17]

F70 Mild Mental Retardation
F71 Moderate Mental Retardation
F72 Severe Mental Retardation
F73 Profound Mental Retardation
F78 Other Mental Retardation
F79 Unspecified Mental Retardation

Characteristics of Subjects with various types of MR [7, 12-14,16]

1. **Mild Retardation (IQ 50-70)**

This is commonest type of mental retardation accounting for 85-90% of all cases. These individuals have minimum retardation in sensory-motor areas.

Pre–school (0-5 years): Overall development is slower than peers. Developmental problems may not be identified until the child starts primary school.

School age (6-15 years): Can master basic learning skills (e.g. reading, writing).Can acquire proper pre-vocational skills.

Adolescence and adulthood (>16 years): Can integrate into community with assistance.

2. **Moderate Retardation (IQ 35-50)**

About 10% of mentally retarded come under this group. The persons with mild & moderate mental retardation can be educated & trained with proper training they can become independent and self sufficient. Their vocational rehabilitation is possible.

Pre–school (0-5 years): Overall development is obviously slower than peers. Can acquire basic communication skills and simple self care abilities.

School age (6-15 years): Can learn some practical skills for daily living. Can live independently to a certain extent in familiar environment and with proper support.

Adolescence and adulthood (>16 years): Can learn to perform simple tasks in specially designed working environment.

3. **Severe Retardation (IQ 20-35)**

Severe mental retardation is often recognized early in life with poor motor development & absent or markedly delayed speech & communication skills.

Profound Retardation (IQ below 20)

This group accounts for 1-2% of all mentally retarded. The achievement of developmental milestones is markedly delayed. They require constant nursing care & supervision**.**

Pre–school (0-5 years): Significant discrepancy in overall development when compared with peers. Some children may also have physical disabilities. Limited communication abilities and response to environment.

School age (6-15 years): Delayed development in motor abilities. Can learn limited communication skills and simple self care tasks.

Adolescence and adulthood (>16 years): Possess simple communication skills. Can master limited basic skills with special support

Clinical characteristics [6]

Children with Mental Retardation usually look like any other children but some may have distinct features like -Small or Large head; Small stature; Protruding Tongue; Blunt features; Drooling; cannot walk with good co- ordination.

Behavioral Characteristics

Brain damage results in skill deficit, which in turn causes challenges in acquiring age, appropriate behaviours, sometimes even manifesting maladaptive behaviors - Slow in response; Unable in making decisions; Difficulty in completing a task uninterrupted even for a short duration; Susceptible to aggressive reaction when demands are not met immediately; Difficulty in remembering; Difficulty in

attending to their self - care needs; Difficulty in complying with group game rules or social norms.

Educational Characteristics

Delay in Development is a characteristic feature such as Slow Reaction; Slow in understanding and learning; Poor attention; Lack of concentration; Short tempered; Poor memory; Lack of co-ordination poor motor development; Slow in speech development.

Oral health among individuals with Mental Retardation [4, 7, 17-20]

Poor oral health can have dramatic effects on an individual's quality of life. In fact, it can cause difficulties with eating, speech impediments, pain, sleep disturbances, missed days of work or school and decreased self-esteem. Individuals with MR, for example, have poorer overall oral health and oral hygiene compared with the general population. The oral health and hygiene of individuals with MR is associated with severity of MR, etiology of MR, residential arrangements and age of the individual.

Dental problems are among the top ten limiting secondary conditions among individuals with MR. One of the most common oral health problems of children and adults with MR is dental caries. Studies, however, do not provide conclusive data on the prevalence of dental caries among those with MR relative to the general population.

Another common oral health problem among children and adults with MR is poor periodontal health and poor oral hygiene with the prevalence estimates of gingivitis being 1.2 to 1.9 times of the general population. Malocclusion, traumatic injuries to teeth, Bruxism and impaired mastication has been also reported in literature.

Barriers to care [7,21]

Despite the high prevalence of health problems among individuals with MR, very little is known about the quantity and quality of services they receive to meet their health needs. Many barriers to care have been cited to explain the low utilization of services and poor quality of care among individuals with MR. The most compelling constraints include uncoordinated systems of health care, providers' lack of training and caregivers', lack of knowledge and abilities, lack of perceived need, inability to express need, and lack of ability for self-care, barriers to accessing and utilizing dental services, poor verbal skills and are restricted in their ability to communicate their needs. Fear and anxiety are the most common barriers to dental care, regular dental attendance; Lack of parental awareness is a major contributory factor for low dental attendance, the knowledge and skills of carers.

Screening and Diagnostic Procedure [12]

(A) Pre-natal Procedures

1. Blood tests.
2. Ultrasonography.
3. Amniocentesis.
4. Foetoscopy.
5. Chorionic Villous Sampling

(B) Neonatal and Post-natal

1. Blood and urine examinations
2. Ultra sonogram.
3. Computerized Tomography
4. Magnetic Resonance Imaging.
5. Ultra Sound
6. Biochemical Tests
7. Electro Encephalography [17]

Treatment [22]

By most definition MR is considered a disability rather than a disease. Currently, there is no cure for an established disability. Although there is no specific medication for MR, many individuals with MR have further medical complications and may take several medications.

Prevention [12]

Prevention of MR is best achieved by avoiding marriages within close relatives with history of Mental Retardation, child bearing between 20- 30 years of age and restriction of family size.

A) Pre natal care: Maintain good maternal nutrition; Exposure to X-Rays should be avoided during first 3 months of the pregnancy; Ensure immunization against

German Measles and Tetanus appropriately: Constant monitoring of high blood pressure; Avoid consumption of alcohol, drugs and tobacco.

B) Natal care: Delivery must be conducted by qualified health professional; immediately after birth the airway must be cleared. In case the baby is blue then he/ she must be put on oxygen immediately

C) Post natal care: Causes need to be detected early for necessary treatment by consulting a qualified Medical Practitioner immediately [17]

Prognosis [9,23]

Most persons with intellectual disability have a life expectancy reaching into adulthood. Persons with intellectual disability who have major malformations and related health problems, more common in those with more severe levels of disability, may have shorter life expectancy based upon these conditions. Life expectancy for those with the more severe levels of intellectual disability is generally less than that for the general population. The decrease is greatest for those who are immobile, have profound intellectual disability, and are unable to feed or care for themselves. For those who were mobile, but non ambulatory, about 50% would be expected to survive to at least 20 years of age. The most common causes of death among individuals with MR are cardiovascular diseases, respiratory illness and neoplastic conditions**.**

Down syndrome

Down syndrome is also known as trisomy 21, trisomy G, and mongolism. It is an easily recognized congenital, autosomal (non sex chromosomes) anomaly characterized by generalized growth deficiency and mental deficiency. Approximately 95% of Down syndrome cases have the extra chromosome 21, making the chromosome count 47 instead of the normal 46. The other 5% are accounted for by other chromosomal abnormalities including translocation (3%) and mosaicism (2%) or partial trisomy. [25]

Incidence [1,24]

→ Down syndrome affects 1 in 600 to 1 in 1000 live births.

→ Down syndrome in India occurs at a rate of 1.4 per 1,000 births. It is estimated that more than 30,000 babies are born with Down's syndrome every year in India.

Types [24]

Three types of chromosomal abnormalities can lead to Down syndrome:

→ Non disjunction - 95% (males 59%, females 41%)

→ Translocation - 4% (females 74%, males 26%)

→ Mosaicism - 1% (may have more subtle features)

Medical complications seem to be similar in all three groups

Manifestations [24,25]

Clinical

Intellectual disability and delayed growth; Vision and hearing problems; Cardiac defects (VSD, ASD, PDA, Tetralogy of Fallot); Characteristic physical features: brachycephalic skull, prominent epicanthic skin folds, small low-set ears, Hypoplasia of mid-facial region; reduced muscle tone, pelvic dysplasia, transverse palmar crease, broad hands and feet, short fingers, and lenticular opacities.

Oral considerations

Lower prevalence of dental caries, Early onset severe periodontal disease (most significant oral health problem, delayed eruption of permanent teeth, congenitally missing and malformed teeth are common. Malocclusion, hypodontia, microdontia, macroglossia, fissured and protruding tongue; tongue thrust, bruxism, clenching, mouth breathing, xerostomia are also reported.

Other Potential Disorders/Concern

Epilepsy; Atlantoaxial instability (fragility of cervical vertebrae/spinal cord); Compromised immune system; Sleep apnea; Increased risk of leukemia; Hypothyroidism, Chronic respiratory infections

Management of Down syndrome [25]

i) Evaluation: Echocardiogram; Ophthalmological assessment; Hearing assessment
ii) Prevention: Obesity; Periodontal disease
iii) Monitoring: Coeliac disease; Thyroid function
iv)Vigilance: Arthritis; Atlantoaxial subluxation; Diabetes mellitus; Leukaemia; Obstructive sleep apnea; Seizures, Sexuality and reproductive health; Dermatological problems; Behaviour problems
v) Medications: The main symptom of Down syndrome patients are seizures and hypertension. Anti convulsants are used to control seizures and calcium channel blockers to alleviate hypertension which has gingival hyperplasia and xerostomia as side effects respectively.

Management in dental office [25]

Many children with Down syndrome can successfully be treated in the dental office

→ Examine patients by the first birthday; monitor tooth eruption patterns and malformations.
→ Give due considerations for associated medical conditions, cardiac status and need for premedication.
→ Care should be taken for increased gag reflex during oral examination.
→ Monitor periodontal disease. Treat as needed and consider specialty referral if indicated
→ Powered toothbrushes may be too stimulating for some children and should be recommended only after determining if the child will tolerate one.
→ Consider prescribing Chlorhexidine or other antimicrobial agents for daily use.
→ Some patients are good candidates for full orthodontic treatment. Maintain primary teeth as long as possible and consider space maintenance and orthodontic consultation for missing teeth
→ Carefully move patients with atlantoaxial instability into the dental chair, giving special attention to the spine and neck. Use pillows to stabilize the patient and increase comfort, as directed by the caregiver
→ Seizure management during treatment: **Remove** all dental instruments from the mouth. **Clear** the area around the dental chair. **Stay** with the child and turn child to one side. **Monitor** airway to reduce risk of aspiration. **Note time** seizure begins: if seizure continues >3 min call **EMS** - Danger of Status Epilepticus (potentially life threatening).

Management Guidance [25]

→ Plan a pre-appointment (in person/ phone) to discuss patient special needs prior to the first visit. Discuss this with the parent or care provider-they know the child best.
→ Schedule their appointments early in the morning.
→ Talk with the parent or caregiver to determine the patient's level of intellectual and functional abilities and explain each procedure at a level the patient can understand.
→ Use short, clear instructions and speak directly to the patient.
→ Minimize distractions, such as sights and sounds, which may make it difficult for the patient to cooperate.
→ Start the oral examination slowly, using only fingers at first. If this is successful, begin using dental instruments.
→ Use the Tell-Show-Do approach when introducing new instruments or procedures.
→ Reward cooperative behavior with positive verbal reinforcement.
→ Develop trust and consistency between the dental staff and the patient. Use the same staff, dental operatory, and appointment time each visit if appropriate

CONCLUSION

A good mental health envisages one's ability to realize one's intellectual and emotional potential. Intellectual disability is caused by many genetic and environmental factors which hampers both cognitive functioning and adaptive behavior. The two

major reasons are Mental retardation and Down syndrome. Individuals with Intellectual disability have plethora of clinical and oral manifestations which makes the diagnosis difficult and so thus management. Due to poor mental conditions along with various unmet barriers these individuals are more at risk of poor oral hygiene which manifests as increase in dental caries and poor periodontal status. Moreover as there is no specific treatment available prevention forms the mainstay of management. The oral treatment of patients with Intellectual disability should thus always be based on concepts of general health .A comprehensive and regular dental care by programme managers, health care professionals should be provided in order to assist maintenance of oral and overall health for this group with special needs.

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