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Management of a Patient with Autism, Dravert Syndrome, Intellectual Disability and Gagging-A Case Study

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INTRODUCTION

This case study discusses the management of a lady with intellectual disability and behavioural difficulty, complicated by gagging issues and a history of seizure.

CASE STUDY

Ms B is a 20-year-old lady, diagnosed with autism and Dravert syndrome with mild intellectual disability. She was currently on clozapine 25mg od, sodium valproate 1000mg bd and clobazam 20mg od. The mother claimed that she would experience a seizure attack about once or twice a year, the last one being one year ago, usually triggered by stress and hunger. Her last dental visit was about 4 years ago, when she was discharged under Paediatric Dentistry care due to her age. After that, she had tried to seek treatment at many different private dental clinics, but she refused to sit on the dental chair and did not cooperate during dental check-up. Ms B lived with her parents. She attended a day care centre for people with disability three times a week. She was the eldest child out of 3. All her siblings and both her parents were fit and healthy. Her hobby was singing and dancing to the songs of her favourite artists, who were Michael Jackson and Britney Spears. She liked to drink cordials as well as eat chocolates and biscuits. She claimed that she brushed her teeth thrice daily, although her mother insisted that she only did it once a day, in the morning. She used an electronic toothbrush and children toothpaste (fluoridated and flavoured). She did not floss.

Upon examination, she presented with incompetent lips and anterior open bite (Figure 1). Her upper and lower lips were cracked, and she claimed to be a mouth breather. She also had caries ICDAS 05 on 12 (palatal), 22 (palatal); ICDAS 03 on 13 (palatal), 37 (buccal), 47 (occlusal, buccal); and ICDAS 02 on 36 (occlusal), 37 (occlusal), 44 (occlusal), 45 (occlusal), 46 (occlusal). There were generalised plaque deposits, as well as generalised gingival inflammation and hypertrophy, with spontaneous bleeding predominantly on buccal of maxillary and mandibular dentition (plaque score 70%, bleeding score 50%).

Saliva testing found that the quality and quantity of her saliva was within normal limits. Radiographic investigation with orthopantomogram showed normal alveolar bone height for maxilla and mandible; missing 48; erupting 18, 28 and 38. Bitewing was not able to be taken due to gagging.

AGREED TREATMENT PLAN/TREATMENT PROVIDED:

1. ***Emergency treatment:***
   - Removal of caries (using hands instruments) and temporary restoration of 12 (palatal) and 22 (palatal).
2. ***Preventative treatment:***
   - Brush two to three times daily with fluoridated toothpaste.
   - Interdental cleaning using floss with handle, once at night after tooth brushing.
   - Dietary advice.
3. ***Definitive treatment:***
   - Periodontal therapy:
     - Mechanical control: plaque removal by professional scaling and polishing and home care (tooth brushing, flossing).
     - Compliance review: 1 week after scaling.
     - Periodontal review: 4 to 6 weeks after scaling.
   - Tooth-coloured (composite) restorations on 12 (palatal), 13 (palatal), 22 (palatal), 23 (palatal), 37 (buccal) and 47 (occlusal, buccal).
   - Preventive resin restoration on 36 (occlusal), 37 (occlusal), 44 (occlusal), 45 (occlusal), 46 (occlusal).

- Fluoride therapy- application of topical fluoride varnish with Duraphat 22600ppm fluoride (5% sodium fluoride) at initial visit and 12th month.

4. Future maintenance:

- Regular review every 3 to 4 months.
- Re-evaluate caries risk at recall visit by conducting saliva testing and dietary analysis.
- Deliver a fluoride varnish treatment at 12th month recall visit.
- Take a bitewing radiograph at 6th month recall visit. (If patient still cannot tolerate intraoral radiograph, consider a new orthopantomogram every 18 months).
- At every recall visit, identify any restorative treatment necessary or the need for thorough professional cleaning as required for patient's periodontal health.

DISCUSSION:

Assessing patients' risk of developing oral disease such as periodontal disease and caries is important in treatment planning and clinical decision-making, reducing requirement for complex periodontal therapy or restorative treatment, improving the outcome of the treatment and reducing financial expenditure on oral health care (1). Ms B presented with a number of management challenges, as discussed below.

1. Medical factors:

a. <u>Autism and intellectual disability</u>

Autism is a neurodevelopmental disorder, characterised by severely impaired social interactions, language, behaviour and cognitive functions (2). Patients with autism were reported to have difficulty communicating and cooperating in the dental office, imposing a challenge to the dental team in providing care (2). About 70% of individuals with autism reported some degree of intellectual disability (2), which may further limit achievement of satisfactory personal and professional oral health care.

Intellectual disability is defined as a condition of having impairments of general mental abilities that can impact on an individual's adaptive functioning in three different domains (3). These domains include conceptual (such as language, reading, writing and memory skills); social (such as empathy, social judgment and interpersonal communication skills); and practical (such as personal care, job responsibilities, money management and recreation). Intellectual disability is diagnosed in individuals where these characteristics begin during the developmental period. To diagnose intellectual disability, the Diagnostic and Statistical Manual of Mental Disorders, DSM-5 emphasizes the need to use clinical assessment based on adaptive functioning (which is the main criteria to determine the severity of impairment), together with an IQ test score of about 70 or below as a standardised testing of intelligence (3). Other commonly used definitions of intellectual disability are the World Health Organisation "ICD-10", Version 2010 (4) and the 2010 classification of America Association on Intellectual and Developmental Disabilities ("AAIDD") (5).

Ms B would be categorised as having mild to moderate intellectual disability. She was able to perform activities of daily living including personal hygiene, laundry, and eating. She was able to read and count slowly up to 20 and follow instruction, and express feelings and desires. However, she had problem understanding conversations and remembering instructions. Ms B was not able to consent for treatment due to her inability to understand (6). In terms of behaviour, Ms B was able to cooperate and sit on the dental chair after being reassured. She was also able to communicate verbally, although slowly, and using simple words and short sentences.

It was important to educate Ms B regarding the importance of oral hygiene as the first step towards establishing non-compromised oral health and to maintain this support. Management of oral disease requires patient's participation to perform self-care procedures, aimed at controlling plaque accumulation and biofilm development (1). Ms B's poor oral hygiene practice may be related to her poor understanding of oral health care and behavioural difficulty, as a result of her autism and intellectual disability (7, 8). It was found that people with intellectual disability had poorer oral

hygiene (9-13) as well as higher prevalence and severity of periodontal disease (14) compared to the general population (15, 16). Individuals with autism were also found to have high prevalence of caries and periodontal disease (17). However, after being taught the proper toothbrushing and flossing techniques, Ms B showed improvement in oral hygiene status, reflected by reduction in plaque score and pocket depth. This suggests that with proper guidance and constant reassurance, people with autism and intellectual disability may still practice good oral hygiene behaviour.

Given her level of intellectual capacity and characteristics of autism, communication poses a significant challenge in delivering effective oral health messages (18). To assist with Ms B's understanding on oral hygiene instruction and dental procedure, simple words and sentences were used. For example, 'gums' instead of gingiva; 'clean your teeth' instead of 'scaling'. Reinforcement in the form of repeated instructions was also performed for Ms B (19). Desensitisation method was applied in formulating her treatment plan, in which the simplest dental procedure was provided at the initial stage, while the more complex treatment was slowly introduced as the patient demonstrated better cooperation (20).

Communication with Ms B was also enhanced by using diagrams such as anatomical pictures of teeth and gingiva, pictures of caries and periodontal disease and illustration to demonstrate the disease process. Other recommended techniques to improve communication with people with autism and intellectual disability include using communication board, electronic communication device, Picture Exchange Communication System (PECS) and facilitated communication, as well as having an interpreter (21, 22).

Oral hygiene instruction was given using her electric tooth brush and employing the tell-show-do technique (20). It is important to have a written record of any communication, including consent and oral hygiene instruction as patient may forget this information. A customised illustrated timetable of daily oral hygiene care was created to help her remember her regimen, and was given together with a leaflet containing oral health information, developed to her level of intellectual functioning.

A thorough explanation was also given in a form of detailed written instruction to her parents as it was also important to educate her carers regarding oral hygiene care (11, 23).

Intraoral radiography and photography was problematic for Ms B due to gagging issue. Her gag reflex was induced by placement of radiographic film/plate in her oral cavity (24). Despite attempting relaxation and distraction technique as proposed by Bassi 2004 on patients with gag reflex, intraoral radiography and photography was still challenging in this case (25). Nevertheless, an attempt in taking intraoral clinical photographs during her second 6th month review visit was successful using systemic desensitisation technique, which was recommended to overcome gagging (25). Relaxation and distraction, via listening to her favourite music from her own mobile device, were also found to be effective in improving her cooperation and tolerance when we performed dental procedures, including scaling and restoration. Other techniques to help overcome gagging include hypnosis, acupuncture and behavioural cognitive therapy, which all require specialised training. The outcomes of these interventions for those with intellectual disability, however remains unclear (25, 26).

b. Dravert syndrome

Dravet syndrome is a severe developmental and epileptic encephalopathy condition (27). The disorder is characterized by the onset of prolonged seizures during infancy, which then progresses to epileptic condition (27). Individuals with Dravert syndrome also present with impaired cognitive, behavioural, and motor functions (27).

Management of Ms B required careful assessment of her epileptic condition. It is important that triggering factors of her developing seizures were identified and avoided. Thorough medical history must be obtained, including the patient's last seizure attack and medication prescribed. Checking patient's compliance to his/her medication is important, as the risk of a seizure attack is potentially high for someone, although with stable epilepsy, who has missed his/her medication for the last 12 to 48 hours (28).

c. Side effects of medications:

One of the side effects of clozapine and clobazam was dry mouth (29). Reduced salivary flow was a recognised factor contributing to caries and periodontal disease, and should be assessed to indicate appropriate management (30) . For Ms B, saliva testing had found that the quality and quantity of her saliva was within normal limits. Her xerostomia may have been related to mouth breathing and an incompetent lip seal. She was advised to maintain adequate hydration and use moisturising gel on her lips to prevent lip cracking (28).

Sodium valproate may cause gingival hyperplasia (29), which may worsen her gingivitis condition. Reinforcement of oral hygiene care was found useful in controlling gingival inflammation in Ms B, without requiring surgical intervention such as gingivectomy or gingivoplasty.

2. Oral risk factors:

Incompetent lips, mouth breathing, anterior open bite

Periodontal disease management in this case was complicated by incompetent lips, mouth breathing habit and anterior open bite, that have been associated as risk factors for periodontal disease. It was found that those with incompetent lip seal showed significantly higher plaque and bleeding index than those with competent lip seal (31, 32). The absence of lip coverage is also thought to diminish the normal cleansing action of saliva, encouraging accumulation of plaque (32). On the other hand, mouth breathing may lead to dehydration of the tissues, impairing their resistance and making them more susceptible to gingivitis (32). In addition, amongst subjects with an incompetent lip seal who were mouth breathers, it was found that gingival index was higher compared to normal breathers (31).Plaque and bleeding scores in the maxillary and mandibular region are also reported to be higher in those with decreased upper lip coverage at rest (31, 33). Plaque accumulation, especially on the anterior region, was also found to be higher in patients with anterior open bite (34).

3. Social factors:

Her dietary habit described high consumption of cariogenic diet and snacking behaviour. Ms B was advised to avoid sucrose and other fermentable carbohydrates in sticky forms or as snacks between meals; limit the intake of acidic drinks containing sucrose and other fermentable carbohydrates; to drink plain water instead of cordial; and to use sugar substitute e.g. xylitol to replace white sugar in her tea. Educating her parents, who are her principal caregivers, as to the importance of a healthy diet was also important.

ORAL HEALTH MANAGEMENT OF MS B

Due to various risk factors as discussed above, Ms B is at high risk of developing periodontal disease and caries. Her treatment focuses on intensive home care regimen and professional care to manage her periodontal disease and maintain her periodontal health, as well as preventing caries and other oral diseases. Her home care regimen consists of mechanical control with tooth brushing (three times daily) and the use of floss with holder. Ms B was also advised to use fluoridated toothpaste every time she performs tooth brushing. Her daily oral hygiene regimen was designed in a manner to accommodate her daily schedule, taking into account any barriers or difficulty. Professional care included weekly appointment at the initial stage to assess her oral hygiene status and practice, with a three to four monthly appointment for maintenance review. A frequent appointment is also helpful to familiarize Ms B with the dental environment in order to overcome her fear and anxiety, help establish rapport and build trusting relationship between herself and the dental team. This may help patient instil positive attitude towards dentistry which in turn improve patients' compliance and determine the success of a designed management plan. Patients who trust and respect their dentist will be more willing to attend dental appointment and be more compliant towards home care regimen (19).

CONCLUSION:

Dental practitioners should take every reasonable measure to ensure safe conduct of dental treatment, especially on patients who have medical complications such as epilepsy. Various modalities in communication strategies and behavioural guidance can be applied during delivery of professional treatment on patients with autism and intellectual disability to improve patients' cooperation. Frequent review may be useful in imparting good oral health knowledge, behaviour and practice in these patients. Educating the patient and carers about oral health care is as important as educating clinicians to provide optimal level of care for these individuals.

CONFLICT OF INTEREST

The author declares no conflict of interest.

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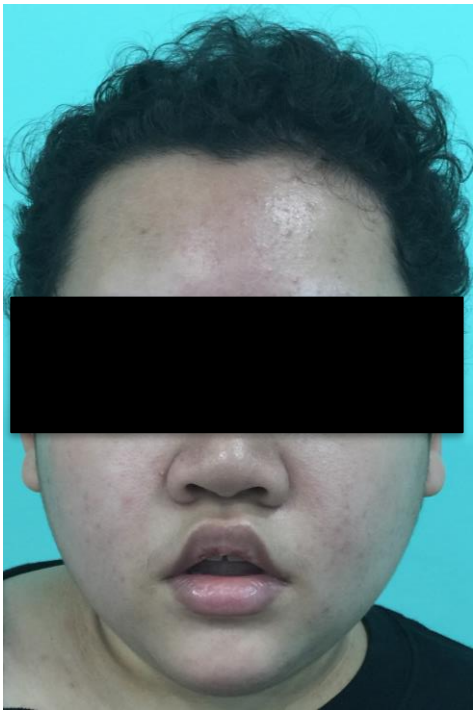
Figure 1: Patients' clinical photographs (only taken at second 6th month review visit due to patients' lack of cooperation and gagging at the initial visits).



Facial view showing incompetent lips. The lips were also dry and cracked.

Frontal view showing anterior open bite

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