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William-Beuren Syndrome: Comprehensive Dental Care in Child Patient

Naila Marzuqi, Aulia Nuur Ainayah,...

Submission








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Naila Marzuqi, Aulia Nuur Ainayah, A...

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Round 1

Round 1 Status

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Notifications

[AMP] Editor Decision	2021-01-27 07:27 AM
[AMP] Editor Decision	2021-02-07 07:34 AM
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Acta Medica Philippina, College of Medicine, University of the Philippines Manila

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tania saskianti (tania_saskianti)

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5. We have rearranged the discussion
6. We already described the figures comprehensively.
7. We have input the parents ethical approval and cardiologist approval
8. We have described William-Beuren Syndrome is a rare genetic disorder with dental manifestation which carried out in one visit and comprehensive with multidisciplinary team .

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Participants

Dr. Abigail Pascual-Domingo (acpascualdomingo)

tania saskianti (tania_saskianti)

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Participants

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tania saskianti (tania_saskianti)

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William-Beuren syndrome: a rare case in Comprehensive Dental Care problems in Child Patient

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Abstract

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The most common symptoms of Williams-Beuren syndrome (WBS) ~~were~~ are heart defects and unusual facial features. The objective of this case report was highlight the preparation of comprehensive dental care of this syndrome. A seven-year-old boy came with multiple dental caries as a chief complaint. Parents reported a history of corrected tetralogy of fallot (ToF) three years ago. Routine medical check-up was carried out annually. Based on the typical signs and symptoms, a diagnosis of William-Beuren Syndrome was made. ~~Comprehensive dental care under general anesthesia on a patient with a history of heart defect and multiple dental caries was the best choice and exhibits a high success rate. Extraction treatment was carried out to avoid the risk of endocarditis infection, restoration is carried out to prevent the progressivity of caries, and space maintainers to restore function and aesthetics.~~

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Keywords: child, dental caries, William-Beuren Syndrome, general anesthesia.

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Introduction

William-Beuren Syndrome (WBS) is a rare genetic syndrome with multisystemic disorder, which has distinctive ~~of~~ physical features, and developmental abnormalities.^[4] The etiology is an autosomal dominant or new mutation. The prevalence is around 1 in 10,000 live births. Based on previous research, Latin America population obtained a prevalence of 55% in boysmale and diagnosed on 11.9 years old, African population obtained a prevalence of 75% in boysmale and diagnosed on 7.7 years old, and Asian obtained a prevalence of 50% in maleboys and diagnosed on 8 years old, but unfortunately there are is has been no researchs showing the specifieregarding the prevalence of our countryin Indonesia.^{[1-4][1,3,4][2]}

Medical manifestations in William-Beuren Syndrome manifestations including intellectual disability, hypersocial behavior, distinctive faciales, short stature, connective tissue anomalies, endocrine abnormalities such as hypercalcemia, and cardiovascular disease (aortic stenosis and peripheral pulmonary stenosis).^[3,5,6]

Craniofacial characteristics of children with William-Beuren syndrome including: a broad forehead, bitemporal narrowing, narrow nasal bridge, periorbital fullness, malar flattening, long philtrum, wide mouth, thick lips vermillion, and prominent earlobes, deficient chin bone and micrognathia and oral manifestation include malformed teeth, enamel hypoplasia, excessive interdental spacing, high arched palate, high prevalence of dental caries.^[7,8]

Dental manifestations in this syndrome are malocclusion, ~~anterior crossbite~~ enamel hypoplasia, malformed teeth, and dental caries. Enamel hypoplasia is the one of dental manifestations in WBS ~~which can~~ increased the risk of dental caries. Since, and the parents focus onf their

child medical treatment on patients it can caused less attention to oral hygiene. —Dental caries are the most common oral disorder. Based on World Health Organization's data, dental caries emerges as one of the oral disorders leading to a person's lower life quality, both aesthetically and functionally^[9].

Dental treatment can be managed with behavior management which serves as one of the techniques employed to treat children with special needs. In a case where non-pharmacological behavior management or pharmacological some treatments, e.g. administering sedatives, does not result in improvement, general anesthesia could be used as a therapeutic option to exercise provide safe and effective dental treatment^[10]. The pediatric dentists can make a dental treatment plans with behavior modification and dental rehabilitation ^[11].

In this patient, because of medical history and treatments needed, dental treatment that aims to prevent and protect psychological development and lower medical risks is the indication of treatment with general anesthesia ^[10-12].

One of the most common Clinical manifestation of includes overriding aorta, right ventricular hypertrophy, and ventricular septal defect ^{[43][14]}. In this patient, because of medical history and treatments needed, dental treatments in medical compromised patient that aims to prevent and protect psychological development and lower medical risks is one of the the indication of treatment with general anesthesia ^[12-14]. General anesthesia is recommended for dental treatment in children with a heart defect, particularly in a multiple dental caries cases. —Dental treatment under general anesthesia forte children with special needs could improve dental treatment quality, particularly preventing children from stress. For children with cyanotic heart defects, dental treatment may lead to stress, which may induce hypoxia, hyperpnea, and irritability. In the present this case, general anesthesia prevents the children's patient's stress from adue to multiple dental visit ^[12,15].

In this case, general anesthesia was performed for dental treatment because somatic and emotional stress, such as pain, fear, excitement, and anxiety caused by the conventional dental treatment can lead cardiovascular exacerbation, and conventional dental therapy was very difficult as a result of the patient's mental retardation and hyperkinesia. ^[16]

Dental treatment on children with heart defects should be exercised with caution, considering such a condition may lead to a risk of infective endocarditis. The treatment should be performed comprehensively by involving pediatric dentist, oral surgeon, cardiologists, anesthesiologists, and pediatrician to determine the treatment procedures, such as to administer antibiotic prophylaxis to prevent infective endocarditis, and improve dental treatment quality ^[15,17].

General anesthesia is recommended for dental treatment in children with a heart defect, particularly in a multiple caries case. Performed dental treatment under general anesthesia to children with special needs could improve dental treatment quality, particularly

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preventing children from stress. On children with acyanotic heart defects, dental treatment may lead to stress, which may induce hypoxia, hyperpnea, and irritability. In the present case, general anesthesia prevent the children's stress from a dental visit^[10,14].

Case Report

A seven-year-old boy visited the Pediatric Dentistry of Universitas Airlangga's Dental Hospital with multiple dental caries as a chief complaint. The parents wanted to have their son's teeth treated to improve his appearance. The boy ~~have~~ has a history of tetralogy of fallot (ToF) three years ago and had been corrected. The patient's mother was infected by Rubella during pregnancy. Based on the typical signs and symptoms, a diagnosis of William-Beuren Syndrome was made by his pediatrician

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(a)

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Figure 1: Facial characteristics (a) broad forehead, epichantal fold, flat nasal bridge, periorbital fullness, malar flattening, smooth philtrum, thick lips vermilion, small chin; (b) large earlobes.

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Intraoral examination showed multiple dental caries. The diagnosis included pulpitis reversible 53,63,71,73,83,84,85, pulpitis irreversible 72,74,81,82, gangrene radix 55,54,52,51,61,62,64,65,75 and post surgery relapse of cleft palate surgery 5 years ago.

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(a)

(a)

(b)

Figure 2. Intraoral examination before treatment (a) the maxilla arch revealed multiple dental caries of 55,54,53,52,51,61,62,63,64, 65 and cleft palate (b) the mandibular arch revealed multiple dental caries of 75,74,73,72,71,81,82,83,84,85. Intraoral examination before treatment (a) Maxilla; (b) Mandibular.

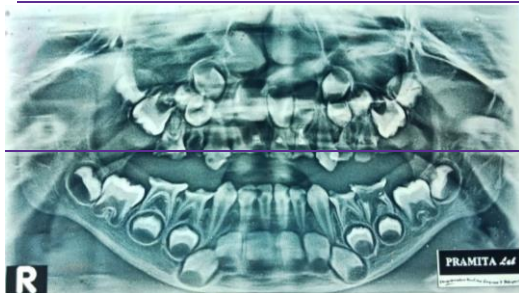
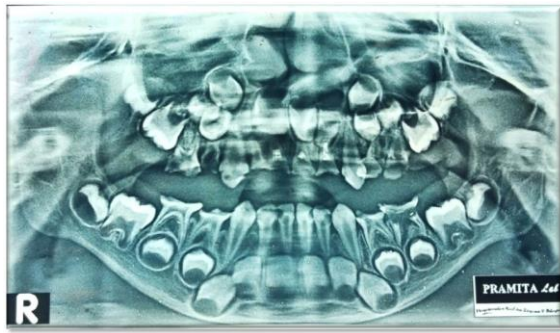


Figure 3: Orthopantomograph examination: tooth buds of permanent teeth, multiple dental caries of maxilla and mandibular arch, and cleft palate. Radiographic Examination

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Case Management

Comprehensive dental care was performed under ~~g~~General anesthesia after patient received the complete examination from the pediatrician, anesthesiologist, and cardiologist. The patient had complete blood count, urine test and ~~admitted to our hospital the day before surgery~~presented in the hospital one day before the general anesthesia treatment.

~~The parents was informed about the treatment procedure, and informed consent was obtained. Dental treatment was performed after obtaining approval from his cardiologist, pediatrician, —and anesthesiologist, cardiologist,~~

The treatment began by administering general anesthesia to the patient, the dental treatment was carried out by a pediatric dentist and oral surgeon. The dental treatment includes ~~of~~GIC restoration on tooth 53,63,71,73,83, occlusal grinding and polishing on tooth 84,85 and application of topical fluoride exercised by the pediatric dentist. Treatment was continued by oral surgeon to extract tooth 55,54,52,51,61,62,64,65,75,72,74,81,82 and suturing. Cleft palate was not corrected according to the parents' informed consent.

Two weeks after the surgery, the patient ~~came visited the oral and maxillofacial department~~to remove the suture. A month after surgery, the patient ~~visited the pediatric dental department and~~was given partial dentures as the space maintainer. Partial denture was fabricated ~~aims~~to improve oral, aesthetic, and mastication functions, thus increasing the patient's self-confidence.



(a)



(b)

Figure 4: 1 month after the surgery, good wound healing (a) the left side: GIC restoration 63,73,71 remain intact; (a) the right side: GIC restoration 53,83 remain intact ~~1 month after the surgery~~



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Figure 5 : Insertion of partial denture as space maintainer (a) the right side ; (b) the left side Insertion of partial denture as space maintainer,

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Discussion

The patient was diagnosed William-Beuren Syndrome, a rare neurodevelopmental disorder that leads to certain clinical, behavioral, and cognitive profiles. It is a multi-systemic condition that is caused by hemizygous deletion of 1.5 to 1.8 Mb on chromosome 7q11.23, ~~containing about 28 genes, which is indicated by a distinctive dysmorphic feature such as various degrees of intellectual disability, congenital heart disease, and abnormal growth pattern.~~^[5,6]

Some manifestation including cardiovascular disease, short stature, (figure 1a) broad forehead, epichantal fold, flat nasal bridge, periorbital fullness, malar flattening, smooth philtrum, thick lips vermillion, small chin (figure 1a), large earlobes (figure 1b), congenital heart disease (Tetralogy of Fallot), and multiple dental caries (figure 2a,b) were seen in this patient but not for intellectual disability.^[3,18]

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~~Facial~~ Enamel hypoplasia is the one of dental manifestation in WBS which can increased the risk of dental caries, and the parents focus of medical treatment on patients can caused less attention to oral hygiene.^[7,8]

~~WBS characteristic that we found in this case include broad forehead, epichantal fold, flat nasal bridge, periorbital fullness, malar flattening, smooth philtrum, thick lips vermillion, small chin, large earlobes (figure 1), and congenital heart disease (Tetralogy of Fallot), and multiple dental caries (figure 2).~~

~~Tetralogy of fallot is a congenital cyanotic heart disease with clinical manifestations includes overriding aorta, right ventricular hypertrophy, and ventricular septal defect.~~^[3,18] [13][14].

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~~In exercising the treatment, a multidisciplinary team comprising cardiologists, oral surgeons, and pediatric dentists was involved. Some dental treatments that were exercised may improve the patient's life quality and self-confidence.~~

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~~In the present case~~ Another case reported, a 28 months old boy diagnosed WBS with cardiovascular disease received dental treatment under pharmacological behaviour management (general anesthesia). Somatic and emotional stress, such as pain, fear, excitement, and anxiety caused by the conventional dental treatment could lead to cardiovascular exacerbation. General anesthesia aims to support a safe, efficient, and effective dental treatment, and to eliminate anxiety, reduce unexpected reflex motions and reaction to the treatment. Besides, it aims to support treating patients with either mental, physical, or medical disorders and to remove pain responses.^[19]

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~~gleado, and conventional dental therapy was very difficult as a result of the patient's mental retardation and hyperkinesia.~~ Curative dental treatment was also necessary because dental caries could pose a risk to endocarditis due to focal infections.^[16]

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A medical evaluation to rule out cardiovascular abnormalities which could affect dental treatment is mandatory. Cases of sudden death have been reported with administration of anesthetic in these individuals. Hence, caution should be exercised while administering anesthesia in these individuals, but in this case performed treatment under general anesthesia was chosen because of ~~there were many treatments that must be done more efficiently in one visit to reduce patient anxiety and increase dental treatment quality.~~^[1]

~~General anesthesia could be applied to patients who are considered non-cooperative due to physical, mental, or medical disorders, young patients, patients with cognitive or emotional immaturity who tend to have extreme anxiety ad requires ext ensive rehabilitation, non-cooperative behaviors, patients requiring extensive treatment, patients who need tooth restoration or surgery that should be performed simultaneously.~~^[12,19,20]

GIC restorations was chosen because of simple handling technique and fluor release (the ion exchanges between glass-ionomer cement and dentine facilitates the remineralization of caries-affected dentine into fluorapatite that provides a caries-resistant). ~~General anesthesia could be applied to patients who are considered non-cooperative due to physical, mental, or medical disorders, young patients, patients with cognitive or emotional immaturity who tend to have extreme anxiety ad requires extensive rehabilitation, non-cooperative behaviors, patients requiring extensive treatment, patients who need tooth restoration or surgery that should be exercised simultaneously, and presence of anatomical variations.~~^[10,19,20]

~~In this case, the patient with medically compromised, and multiple dental treatments are recommended performed under general anesthesia. Based on the medical history, the patient hasve a history of tetralogy of fallot (ToF). Root canal treatment was not recommended in this case for deciduous teeth with a bad prognosis due to high incidence of chronic infections. Tooth extraction— ended with suturing to ensure wound healing was performed and followed by removable partial denture insertion. The utilization of sutures in attempting primary wound closure and the role of suturing in providing wound stabilization.~~^[21,22]

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~~The patient was given r~~Removable partial denture as a space maintainer ~~was fabricated that aims to maintain the space of the dental arch, fill the physiological space, and guidance of the permanent tooth eruption.~~^[23] ~~(alasan suturing ended with suturing to ensure wound healing) cari sitasi~~

~~Treatment was carried out in one visit and comprehensive with multidisciplinary team (cardiologist, anesthesiologist, oral surgeon, pediatric dentist, and pediatrician). EThe treatment include extraction, restoration, occlusal grinding was performed in this case. Because there~~

~~are many treatments that must be done more efficiently in one visit to reduce patient anxiety and increase dental treatment quality. It can reduce the progression of caries, improve aesthetics, function and increase self-confidence.~~

Conclusion

Regarding various consideration, comprehensive dental treatment for William-Beuren syndrome in one visit was deemed suitable. General ~~a~~ anesthesia was chosen as appropriate behavior management pharmacologically. ~~In this case, general anesthesia facilitated a safe, effective dental treatment that minimizes the risk of complication on the patient and lowers the patient parents' anxiety. Comprehensive care is the best option to make a treatment plans in patient with William-Beuren syndrome.~~

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References

1. Wong D, Ramachandra, Sulugodu S, Singh AK. Dental management of patient with Williams Syndrome - A case report. 2015.
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Messages

Note	From
<p>Dear Dr. Saskianti,</p> <p>Please see the attached file galley proof - William-Beuren Syndrome: Comprehensive Dental Care in Child Patient.</p> <p>For your approval.</p> <p>If you have corrections, please message us ASAP.</p> <p>Maraming salamat po.</p> <p>Sincerely,</p> <p>Arianne Patana Acta Medica Philippina ALPATANA@UP.EDU.PH</p> <p> mcardorniga, 050621_AM (in press)_CR-13_[2725]_5pp (William-Beuren Syndrome).pdf</p>	<p>arianne_galley 2021-05-06 07:21 AM</p>
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