

BEHAVIORAL MANAGEMENT APPROACHES IN DENTISTRY AND STRESS MANAGEMENT FOR A PATIENT WITH SALT-WASTING CONGENITAL ADRENAL HYPERPLASIA AND AUTISM SPECTRUM DISORDER: A CASE REPORT

Manejo conductual en odontología y control del estrés en paciente con hiperplasia suprarrenal congénita con pérdida de sal y trastorno del espectro autista: Reporte de caso

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ABSTRACT

Introduction: Congenital Adrenal Hyperplasia (CAH) refers to a group of autosomal recessive disorders characterized by a deficiency that prevents or hinders cortisol biosynthesis. The most frequent and severe form is the salt-wasting type, which involves a significant decrease in cortisol and aldosterone levels resulting in potentially fatal shock due to insufficient cortisol response to stress. On the other hand, Autism Spectrum Disorder (ASD) is one of the most prevalent and complex neurodevelopmental disorders. It is characterized by difficulties in communication and social interaction, and by restricted and repetitive patterns of behavior, interests, and activities.

Objective: To present a case of dental treatment in which behavioral management was used to reduce stress in a patient with salt-wasting CAH and ASD, as an alternative to pharmacological management.

Case Report: This report presents the case of an 11-year-old male patient diagnosed with salt-wasting CAH and Autism Spectrum Disorder (ASD). Reason for consultation: the patient's parents requested an alternative treatment for caries that would not require general anesthesia. The patient shows negative behavior towards dental care. The parents reported the inability to manage his son's behavior during a previous dental experience in an operating room under general anesthesia. The patient was treated at the Special Patients Clinic, Faculty of Dentistry, Universidad de Concepción, Chile, between the years 2015 and 2023. Restorative and preventive dental treatment was planned. The frequency of care varied from once a week to every two weeks during the adaptation and restorative stage; and every three months in the preventive and maintenance stage. The treatment was carried out using behavioral modeling strategies in dental care as an alternative to pharmacological management for stress control. In the initial interventions, a negative behavior was observed. However, after a year of dental treatment involving psychoeducational techniques, the patient gradually progressed to a definitely positive behavior.

Conclusion: This case shows that the use of psychoeducational techniques and strategies in dental treatment was a beneficial and effective alternative to pharmacological methods to manage stress and medical crises. An improvement in the patient's behavior was achieved throughout the course of treatment.

Keywords: *Autism spectrum disorder; Adrenal hyperplasia, Congenital; Behavior; Dentistry; Patients; Child*

RESUMEN

Introducción: La Hiperplasia Suprarrenal Congénita (HSC) corresponde a un grupo de trastornos autosómicos recesivos. Su déficit impide o dificulta la biosíntesis de cortisol. La forma más frecuente y grave es aquella con pérdida de sal, involucra una disminución importante de cortisol y aldosterona con resultado de shock potencialmente mortal por respuesta insuficiente del cortisol al estrés. Por otra parte, el Trastorno del Espectro Autista (TEA) es uno de los trastornos del neurodesarrollo más frecuentes y complejos, se caracteriza por dificultades en la comunicación e interacción social y por patrones restringidos y repetitivos en el comportamiento, intereses y actividades. **Objetivo:** Presentar un caso de tratamiento odontológico basado en manejo conductual para evitar el estrés en un paciente con HSC con pérdida de sal y TEA como alternativa al manejo farmacológico.

Reporte de Caso: Se reporta el caso de un paciente masculino de 11 años con diagnóstico de HSC con Pérdida de Sal y Trastorno del Espectro Autista (TEA). Motivo de consulta: los padres solicitan una alternativa de tratamiento para caries, sin anestesia general. El paciente tiene una conducta negativa frente a la atención odontológica, se relata experiencia dental previa en pabellón con anestesia general por imposibilidad de manejo conductual. Fue atendido en la Clínica de la Unidad de Pacientes Especiales, Facultad de Odontología, Universidad de Concepción, Chile, entre los años 2015 y 2023. Se planificó realizar tratamiento odontológico restaurador y preventivo con una frecuencia de atención que varía de una vez por semana a cada dos semanas en la etapa de adaptación y restauradora; cada tres meses en etapa preventiva y de mantención. El tratamiento se llevó a cabo mediante técnicas de manejo conductual en la atención odontológica como alternativa al manejo farmacológico para el control del estrés. En las primeras intervenciones se observó una conducta negativa. Luego de un año de tratamiento odontológico con técnicas psicoeducativas fue paulatinamente progresando a una conducta definitivamente positiva.

Conclusión: En este caso, el uso de técnicas psicoeducativas en el tratamiento odontológico fueron una alternativa beneficiosa y efectiva para evitar el estrés y crisis médica, en contraposición al manejo del estrés mediante farmacología. Se logró una progresión favorable en la conducta del paciente a lo largo del tratamiento.

Palabras Clave: *Trastorno del espectro autista; Hiperplasia suprarrenal congénita; Conducta; Odontología; Pacientes; Niño.*

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INTRODUCTION

Congenital Adrenal Hyperplasia (CAH) is a group of autosomal recessive disorders caused by genetic mutations of different enzymes involved in the adrenal steroidogenesis pathway. These deficiencies prevent or hinder the biosynthesis of cortisol.¹⁻³ It is usually the result of mutations or deletions of the CYP21B gene and the pseudogene CYP21A,⁴ which produces a deficiency of the enzyme 21-hydroxylase in 90%-95% of cases.^{3,5}

The classic form of CAH due to 21-hydroxylase deficiency is the most common, with an estimated prevalence of 1/14,000, which is why it is considered a rare disease.⁶ The hypothalamus and pituitary gland react to low cortisol levels by increasing the secretion of corticotropin-releasing hormone and adrenocorticotrophic hormone, respectively. This stimulation induces hyperplasia of the adrenal cortex, increasing steroid synthesis mediated by the enzymes 21-hydroxylase, 11-beta-hydroxylase, and 17-alpha-hydroxylase. Deficiency of these enzymes disrupts steroid production, causing the accumulation of precursors instead of cortisol, leading to the production of androgens.^{1,3}

The condition presents a spectrum of clinical manifestations, which are classified into two forms: classic (salt-wasting and simple virilizing) and non-classic (symptomatic and cryptic).⁷ In the salt-wasting form (SW), which is the classic and most severe expression of the disease, there is a significant cortisol deficit, which is practically absent, and a reduction in aldosterone. This manifests in both sexes as severe acute salt-wasting crises during the neonatal period and in a delay in growth.^{6,8,9}

Without appropriate treatment, affected individuals may experience critical situations known as "*adrenal crisis*" and require urgent treatment.¹⁰ Clinical manifestations depend on the degree of cortisol and aldosterone deficiency.¹¹ Adrenal insufficiency crises have significant morbidity and mortality if patients do not receive adequate and prompt treatment.² In addition, these crises may cause neurological developmental impairment and increase the risk of medical emergencies,

triggering a potentially fatal shock due to hypoglycemia, hyponatremia, hypovolemia, acidosis, and hypotension.⁶

Other observed manifestations may be hyperkalemia, polyuria, dehydration, decreased growth, and hyperpigmentation.⁹ CAH is a chronic disease that requires continuous monitoring and intervention, involving multiple medical specialties and demanding a high level of commitment from the patients and their parents.¹

On the other hand, Autism Spectrum Disorder is one of the most frequent and complex neurodevelopmental disorders, characterized by difficulties in communication and social interaction and by restricted and repetitive patterns of behavior, interests, and activities.^{12,13} Dental care can be particularly challenging for a person with ASD, mainly due to the multiple sensory alterations that can trigger sensory dysregulation,¹⁴⁻¹⁶ which, in turn, can lead to uncooperative behavior.^{17,18}

The Frankl Scale^{18,19} is commonly used to assess the behavior of patients with ASD. This scale includes four categories (Annex 1):

1. Definitely Negative
2. Negative
3. Positive
4. Definitely positive

Given the challenges of providing dental care, a high percentage of patients are frequently treated under general anesthesia. However, there are psychoeducational techniques and strategies that facilitate dental procedures and can help reduce excessive use of general anesthesia in these patients.²⁰

The present case is more complex due to the presence of both conditions: Salt-wasting CAH and ASD. Dental management of patients with CAH involves outpatient treatment, supplemented by preventive pharmacological management of stress using prednisone administration protocols, with dose adjustments or doubling the dosage before and after the intervention.^{11,21} In other cases, dental treatment under general anesthesia has been reported combined with corticosteroid dose adjustments.²²

Dental care can be a significant stress factor for patients with CAH. Therefore, it is imperative for a dentist to recognize the signs of an acute adrenal crisis and, even more so, prevent it from occurring. There are several strategies that can be implemented to achieve patient adaptation to the clinical environment, procedures, and dental staff.²³

A case of outpatient dental care is reported, involving behavioral management using desensitization techniques to perform preventive treatments and extractions. However, the restorative stage was ultimately carried out under general anesthesia.²⁴

The importance of reporting this case lies in the fact that there are few reports of patients with this condition, and it is even less common to find cases in which the complete dental treatment is performed without the use of general anesthesia. In addition, stress management in these patients is generally described using pharmacology or interventions in the operating room. In contrast, the present case was managed using psychoeducational techniques.

The aim of this case report is to present a dental treatment approach based on behavioral management to prevent stress in a patient with salt-wasting CAH and ASD, as an alternative to the most frequently used pharmacological management.

CASE REPORT

An 11-year-old male patient, diagnosed with salt-wasting CAH and ASD, under dental treatment from the age of 5 years old at the Clinica de la Unidad de Pacientes Especiales, Faculty of Dentistry, Universidad de Concepción, Chile, between the years 2015 and 2023. His parents report that their reason for consultation is requesting an alternative treatment for caries that does not involve the use of general anesthesia. The patient's relevant medical history includes full-term pregnancy with cesarean delivery.

At six months of age, the patient suffered cardiorespiratory arrest and was placed in an induced coma for 15 days. He presents accelerated bone development, hypotonia, hyperlaxity, acropachy (clubbed fingers), thumb atrophy, and genu valgum (Figure 1 C). Additionally, he has delayed language development, having begun speaking at the age of 5. While his receptive language skills enable him to understand simple commands, his expressive language abilities are significantly impaired.

Family medical history includes no relevant conditions for the patient's mother; his father has insulin resistance. Maternal and paternal grandparents have medical history of arterial hypertension and diabetes mellitus. The patient's current medications and dosage are

Figure 1. Photographs of the patient at 5 and 6 years of age.



A: Patient's front photographs at 5 years old.

B: Patient's profile photographs at 5 years old.

C: Patient's clubbed fingers at 6 years old.

Figure 2. First dental care provided to the patient at 5 years old.



Process of applying psychoeducational techniques for behavioral management.

Figure 3. Photographs of patient's upper and lower arches, at 6 years old.



A: Photograph of the upper arch (tooth 6.2 is missing).

B: Photograph of the lower arch.

prednisone 3 mL once a day, fludrocortisone 0.1 mg twice a day, anastrozole 1 mg once a day, Decapeptyl® (triptorelin) 3.75 mg once every 3 months.

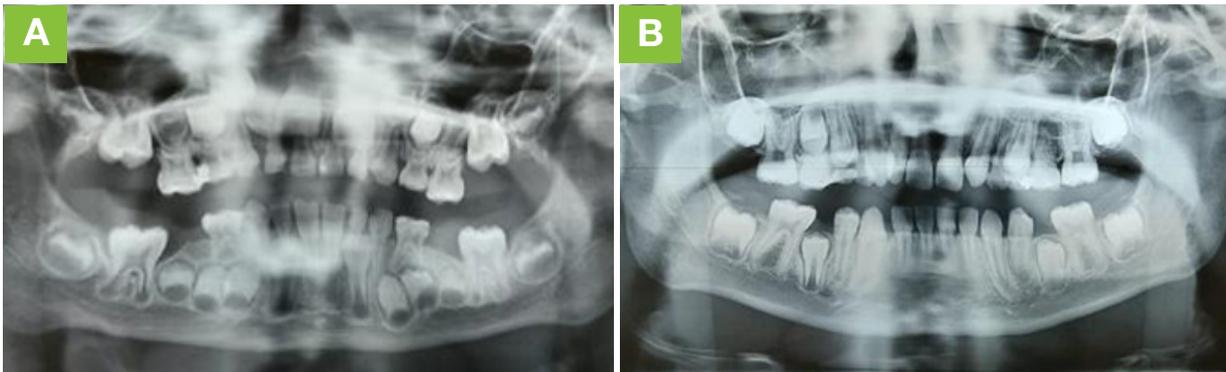
Panoramic radiography showed agenesis of teeth 2.2, 2.5, 1.8 and 2.8. Extraoral clinical examination revealed angular cheilitis and labial incompetence (Figure 1A and 1B). On intraoral examination, temporary dentition, multiple proximal carious lesions of enamel and dentin were observed in teeth 5.4, 5.5, 6.1, 6.5, 7.4, 8.4, as well as the loss of teeth 7.5 and 8.5 (dmft= 8), and absence of tooth 6.2.

The Simplified Oral Hygiene Index was 0.83 (good). Other findings include deep palate, increased overbite, and worn enamel on facets. The caregivers report oral hygiene

habits that include assisted tooth brushing twice a day with a manual toothbrush, toothpaste, and mouthwash. The patient has parafunctional habits, such as using a bottle or pacifier at night, and exhibits (mixed) mouth breathing.

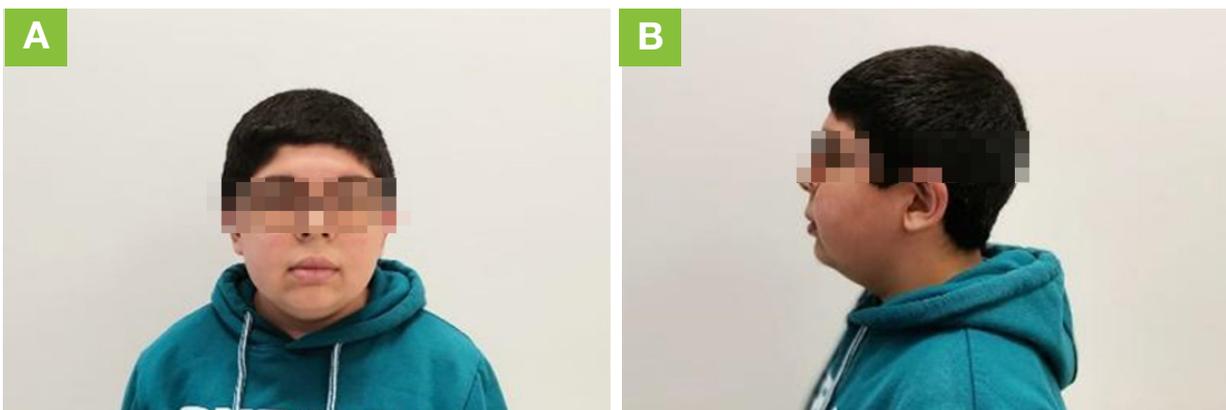
Regarding behavioral history, the patient does not exhibit any self-injurious or other-directed injurious behaviors. His dental behavioral history shows negative behavior towards dental care. His previous dental experience was exclusively in an operating room under general anesthesia, with the aim of extracting teeth 7.5 and 8.5 due to the inability of managing his behavior in the clinical setting. Informed consent was obtained from the caregivers who authorized the publication of this clinical case report.

Figure 4. Panoramic radiographs taken during dental treatment in 2017 and 2021.



A: Panoramic radiograph, agenesis of teeth 2.2 and 2.5.
B: Panoramic radiograph, year 2021.

Figure 5. Photographs of the patient at 11 years old, during his last check-up.



A: Patient's front photographs at 11 years old.
B: Patient's profile photographs at 11 years old.

Figure 6. Intraoral photographs of the patient's last check-up, at 11 years old.



A: Intraoral photograph of the upper arch (teeth 2.2 and 2.5 are missing).
B: Intraoral photograph of the lower arch.

Table 1. Dental care schedule by milestones, age, behavioral management techniques, and behavior assessed using the Frankl Scale.

Milestones	Date (dd-mm-yy)	Age	Behavioral management	Frankl scale (behavior)
First consultation	22/03/2017	5 years	Successive approaches, Anticipation, Desensitization Positive reinforcement	2
First restoration with Atraumatic Restorative Treatment (ART). (Figure 2)	05/04/2017	5 years	Tell-Show-Feel-Do (T-S-F-D), Distraction, Voice control, Desensitization, Positive reinforcement	2
First local anesthesia	03/05/2017	5 years	Distraction, Positive reinforcement	3
Suspension of dental treatment due to hospitalization (medical crisis)	14/06/2017	5 years	-----	---
Patients resumes treatment	05/07/2017	5 years	Successive approaches, T-S-F-D, Voice control, Desensitization, Positive reinforcement	2
Space maintainer cementation	09/08/2017	5 years	T-S-F-D, Desensitization, Positive reinforcement	2
Resin restorations with absolute isolation (Figure 2)	25/04/2018	6 years	T-S-F-D, Desensitization, Positive reinforcement	3
Preventive phase check-up (Figure3)	25/08/2018	6 years	Anticipation, T-S-F-D, Social positive reinforcement	4
Lingual Arch Cementation	26/12/2018	7 years	Desensitization, Positive reinforcement	3
Monitoring and X-rays during the SARS-CoV-2 pandemic (Figure 4)	30/07/2021	9 years	Anticipation, Positive reinforcement	4
Preventive treatment	15/10/2021	10 years	T-S-F-D, Positive reinforcement	3
The patient is referred for orthodontic treatment at another health facility.	08/04/2022	11 years	Positive reinforcement	4
Check-up	02/11/2022	11 years	Positive reinforcement	4
Check-up (Figure 5)	24/03/2023	11 years	Positive reinforcement	4

RESULTS

A dental care schedule was prepared by milestones, age, behavioral management techniques, and behavior assessed using the Frankl Scale (Annex 1).

In this case, a non-pharmacological intervention was implemented to control stress during dental treatment, with the consent of the caregivers (parents). This intervention was based on the psychoeducational techniques described below:

Successive approaches

Gradually introducing the patient to dental care, starting with the least invasive procedures and progressing to the most invasive ones, while reinforcing each activity. This technique was primarily used when starting a new dental procedure.

Tell-Show-Feel-Do (T-S-F-D)

This technique consists of helping the patient understand the procedures to be performed.

- **Tell:** Explain, using a communication method understandable by the patient, what is going to be done.

- **Show:** demonstrate on a model or person what will be done.
- **Feel:** Allow the patient to experience visually, tactilely, olfactorily, gustatorily, and auditorily the stimuli that will be felt during the procedure.
- **Do:** Perform the planned procedure. This technique was mainly used for more invasive procedures, such as the use of rotary instruments and space maintainer treatment.

Positive reinforcement

Reinforcing positive behavior that follows a stimulus, making it stronger and more likely to be repeated in similar situations. Reinforcers were identified in the anamnesis during the clinical history. Activity reinforcers (allowing him to play with small cars) and social reinforcers (congratulating him and praising his good behavior) were used primarily.

Anticipation

Moving forward in time in the actions to be carried out. It was used to familiarize the patient with a new procedure.

Desensitization

Exposing the patient to a series of experiences presented in an order that evokes anxiety, progressing only when the previous one is accepted in a relaxed state. This technique was used transversally throughout the treatment. During each procedure, the patient's anxiety levels and behavior were assessed, stopping clinical procedures at any sign of stress.

Voice control

controlled change in the tone, rhythm, volume, and firmness of the voice. It is used to capture the patient's attention when procedures are interrupted by his disruptive behavior, and to encourage and reinforce a desired behavior.

Distraction

Diverting attention from harmful stimuli and modulating perception and expectations. It helps to cope with short-term stressors. This technique was used when performing anesthetic techniques.

DISCUSSION

Dental management of CAH is documented in a small number of case reports. On the one hand, outpatient dental treatment protocols have been described involving preventive pharmacological management of stress through prednisone administration protocols, with dose adjustments or doubling of the dose before and after the intervention.^{11,21} Additionally, dental treatment under general anesthesia combined with corticosteroid dose adjustments has also been reported.^{11,21,22}

On the other hand, the literature describes alternatives to behavior control approaches for difficult-to-manage patients. There are some psychoeducational techniques and strategies that facilitate dental procedures, such as oral examination, which can help reduce the excessive use of general anesthesia with these patients.²⁰ In this case report, we used this approach to manage and prevent episodes of stress, serving as an alternative to pharmacological management and intervention under general anesthesia. This approach was initially requested by the caregivers, to whom the strategy was explained. After that, they provided their consent.

The patient had previously received dental treatment under general anesthesia. However, after that intervention, dental treatment became necessary again over time. Although behavioral modeling requires more prolonged care, it has proven beneficial, since it has allowed the patient to receive frequent dental care over the long term. This contrasts with the use of general anesthesia, which carries the inherent risks associated with surgical procedures in a clinical setting.

Constant monitoring of behavior was carried out during each procedure, which was crucial in preventing a stress episode. When a definitely negative behavior was identified (Frankl Scale = 1), the clinical procedure was stopped immediately to avoid a crisis. The relevant psychoeducational techniques and strategies to reduce the stress level and encourage the patient's cooperation were used. Once a procedure was completed without causing negative behavior, the patient progressed to the next stage for another more complex procedure.

In the initial interventions, negative behavior was observed (Frankl Scale=2). During the course of treatment, there was a progression to positive behavior (Frankl Scale=3).

After one year of dental treatment using psychoeducational techniques, a definitively positive behavior was achieved (Frankl Scale=4). Dental treatments were performed without stress and without the need for pharmacological control or care in an operating room, therefore reducing the medical risk for the patient. This approach allowed for frequent preventive check-ups and more complex treatments, such as orthodontic therapy, which would not have been possible without the use of the management techniques described.

CONCLUSION

Behavior management techniques proved to be effective during dental treatment in preventing stressful situations and medical crises. They served as a beneficial and effective alternative to the use of medications. An optimal level of cooperation and comfort for the patient was achieved, meeting the caregivers' expectations to avoid dental treatment under general anesthesia and its associated risks.

It is important to consider that the patient's behavior is closely related to his or her stress level, making constant monitoring of behavior essential to preventing a medical crisis caused by stress. In this case, a clear improvement in behavior was observed over time, enabling preventive, restorative and more complex treatments to be completed successfully. Both the patient and the caregivers said that they were satisfied with the treatment outcomes.

Annex 1. Frankl behavioral scale.

Score	Skoring	Observed behavior
1.	Definitely positive	Good rapport with the dentist, interested in the dental procedures, laughing and enjoying the situation.
2.	Positive	Acceptance of treatment; at times cautious, willingness to comply with the dentist, at times with reservation but patient follows the dentist's directions cooperatively.
3.	Negative	Reluctant to accept treatment; uncooperative, some evidence of negative attitude but not pronounced, i.e. /sullen, withdrawn.
4.	Definitely negative	Refusal of treatment, crying forcefully, fearful or any other overt evidence of extreme negativism.

CONFLICT OF INTERESTS

The authors declare no conflicts of interest.

ETHICS APPROVAL

Informed consent was granted by the caregiver (mother of the patient).

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AUTHORS' CONTRIBUTIONS

Cecilia Cantero-Fuentealba: Conceptualization; Investigation; Writing - Original Draft; Writing- Review and editing.

Linda Henríquez-González: Investigation; Writing - original Draft; Writing- Review and editing.

Verónica Cabezas-Osorio: Writing- Review and editing.

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PEER REVIEW

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PLAGIARISM SOFTWARE

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