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CASE REPORT

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Rehabilitation of ectodermal dysplasia patient with a telescopic denture in the maxilla and mandibular implant assisted overdenture: A case report

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Abstract

Hypohidrotic ectodermal dysplasia is a heritable disease, characterized by hypodontia, hypotrichosis, and anhidrosis. This clinical report demonstrates prosthetic rehabilitation of a patient complaining of impaired mastication and the odd appearance of her jaws. Maxillary telescopic overdenture and implant-supported mandibular prostheses were fabricated to improve mastication, speech, and esthetics of the patient.

KEYWORDS

dental implant, ectodermal dysplasia, hypodontia, removable prosthodontics, telescopic crown

1 | INTRODUCTION

The ectodermal dysplasias (EDs) are a heterogeneous group of disorders that affect ectodermal structures, which involve hair, teeth, nails, and sweat glands.[1] In 1929, the expression of hereditary ectodermal dysplasia was found by Weech, who suggested the term anhidrotic for those with inability to sweat,[2] which was changed by Felsher in 1944, to become hypohidrotic, because the author did not agree with the idea of being hypohidrotic patients do not have any sweat glands.[3] Freire-Maia defined (ED) as a syndrome characterized by two or more of the following traits:

(i)Trichodysplasia (abnormal hair).
(ii) Anodontia or oligodontia (abnormal or missing teeth).
(iii)Onchodysplasia (abnormal nails).
(iv) Dyshidrosis (abnormal or missing sweat glands).[4]

Hypohidrotic ectodermal dysplasia is 1 out of 150 types of ectodermal dysplasia in humans.[5] It is also known as "Christ–Siemens–Touraine" syndrome. It has a high frequency, with a prevalence of 1 per 100,000 births. The triad of hypotrichosis, hypohidrosis, and hypodontia considered as diagnostic criteria.[4,6,7]

1.1 | Genetic overview of hypohidrotic ectodermal dysplasia

It has a variety of models of inheritance. The etiology in most cases is mutations in the EDA (Ectodysplasin A) gene, inherited in an X-linked recessive pattern, in which, exhibited clearly in males who are affected much more than females carrying this gene. On the contrary, fathers cannot pass X-linked traits to their sons[5] except if the child has XXY chromosomes (Klinefelter's syndrome), then the father could have given an X and a Y and the one X could have come from the mother.[8] Bearing in mind that the mutations could happen spontaneously without any family history of this syndrome, in which case a *de novo* mutation would have occurred. Mutations in the EDA, EDAR,

and EDARADD genes cause HED. EDA is the only gene known to be associated with X-linked HED (XLHED). 95% of individuals with HED have the X-linked form. The genes EDAR and EDARADD are known to be associated with both autosomal-dominant and autosomal-recessive forms of HED. Mutations in these genes account for 5% of HED.[9] Any ectodermal derivative may be affected in EDs.[10]

1.2 | Clinical features

Reduced number of sweat glands in HED patients leads to a decrease in the ability to sweat (hypohidrosis), which may lead to a dangerous increase in the body temperature.[5] In turn, it may cause cerebral damage or, uncommonly, death due to hyperthermia.[11]

Sparse scalp and body hair (hypotrichosis) is one of the main clinical features, whereas the hair of the affected patient tends to be light-colored, brittle, and slow-growing.[5]

Oral manifestations involve the number and morphology of teeth, growth of the maxilla, the mandible, and the quantity of saliva. Anomalies in teeth number can be divided into three groups:

- Hypodontia (absence of fewer than six teeth).
- Oligodontia (absence of six or more teeth).
- Anodontia (complete absence of teeth).[12]

A prominent forehead, thick lips, and a flattened bridge of the nose are the main facial features. In addition, thin, wrinkled, and hyper-pigmented skin around the eyes, as well as, chronic skin problems such as eczema may be noticed,[5] and convex nails.[13]

Morphology varies between individuals and families. Most forms of ED are not associated with mental retardation. Dental problems vary, depending on the pattern of ED, from widely spaced teeth to poorly shaped crowns and roots, defective enamel formation, malformed teeth, and partial or complete anodontia.[1]

Congenitally missed teeth are associated with failure of normal development of the alveolar process.[14,15] Most patients present with class III maxillomandibular relationships, due to underdevelopment of the maxilla compared to the mandible.[1]

1.3 | Treatment protocols

Growth of the middle and lower third of the face get improved by removable prosthetic and orthopedic appliances, in comparison with untreated HEDs.[16]

Permanent teeth are found in most ED patients, often in the incisor and first molar areas. They tend to be small and abnormally shaped. We should avoid extraction, whereas they can be restored with composite resin crowns or used as overdenture abutments. Implants also can be used in this case. The final decision of retaining teeth depends on root and crown morphology, the position in the arch, and the type of prosthetic treatment. Many patients suffer from reduced VDO (vertical dimension of occlusion) and deficiency in lip support, which can be restored with either implant-supported fixed or removable prostheses.[1]

2 | CASE REPORT

This is a case report of a 22-year-old female patient, was referred from a general dentist to the department of prosthodontics, Damascus University, with a complaint of abnormal and unpleasant appearance of her edentulous jaws and lack of chewing ability.

2.1 | Medical and family history

Her parents were normal with consanguineous marriage. One of her siblings was diagnosed with ectodermal dysplasia. Her height was 158 cm, and she weighed 48 Kg. Her mental development was within normal limits, and she had a normal educational status.

There was no significant medical history.

2.2 | Clinical manifestations

The patient presented with dry and pale skin, hyperpigmentation and linear wrinkles around the eyes, protuberant lips, and a depressed nasal bridge. In addition, she had a prominent chin resulting in a concave facial profile, midface reduction, and an aged appearance. The patient had a severe intolerance to heat, which may be attributed to underdeveloped sweat glands. She has suffered from eczema and has been treated with skin rejuvenating creams (Figure 1).

An intraoral examination revealed a severe atrophic mandibular ridge with minimal height and width, as well as, severe hypodontia in the maxilla (she only had permanent conical canines and primary first molars in each side of the maxilla with anodontia in the mandible (Figure 2).

The clinical findings were confirmed by a panoramic radiograph (Figure 3).

According to her clinical features, she was diagnosed with hypohidrotic ectodermal dysplasia syndrome.

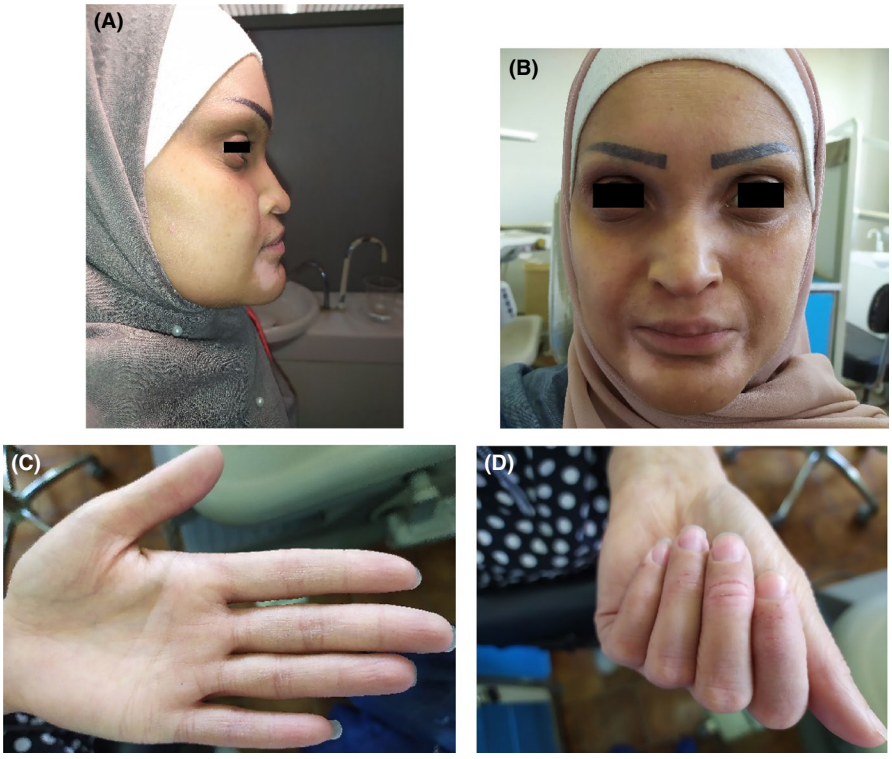


FIGURE 1 (A) Clinical lateral preoperative view showing a depressed nasal bridge, prominent chin, and concave facial profile. (B) Frontal view showing hyperpigmentation and linear wrinkles around the eyes. (c) Palmer Hyperkeratosis. (D) Eczema

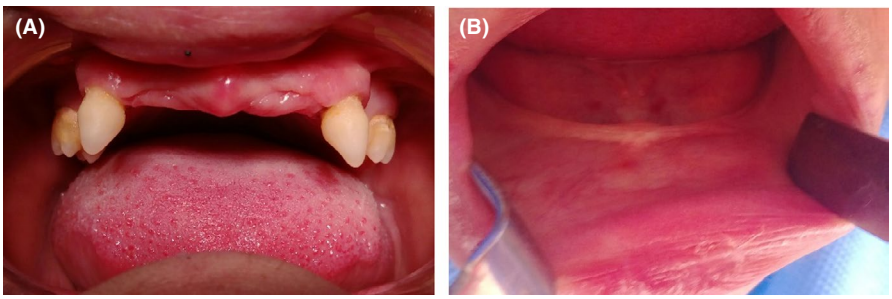


FIGURE 2 (A) Patient presented with only 4 teeth (2 canines and 2 first molars), (B) Mandibular oligodontia was presented

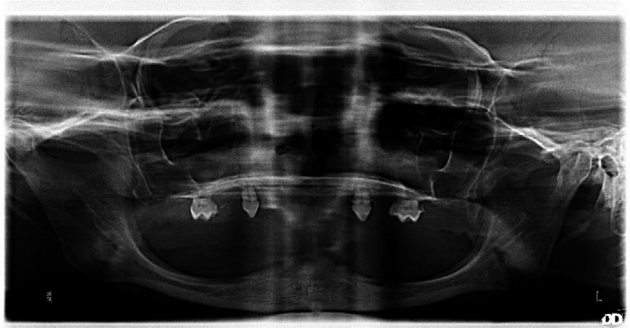


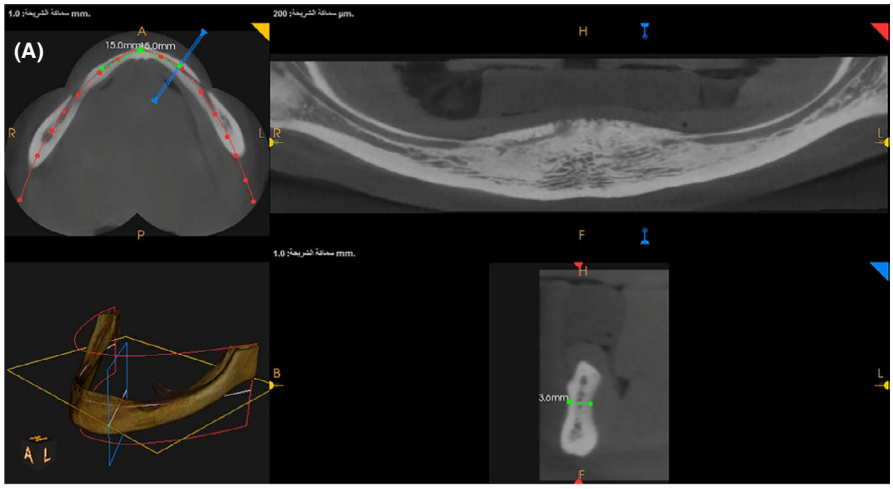
FIGURE 3 Panoramic radiograph shows significant underdevelopment of mandibular ridge and oligodontia in the maxilla

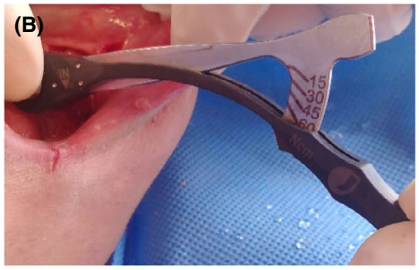
2.3 | Management

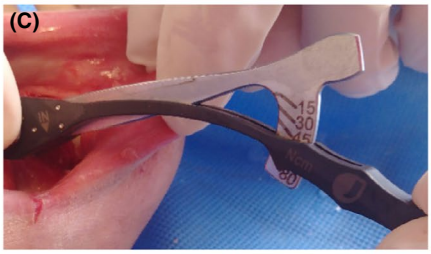
Possible treatment plans and difficulties were discussed with the patient. The final treatment plan was decided as dental implants in the anterior part of the mandible, and telescopic overdenture in the maxilla, as in this type of retainers, the denture can easily be retrieved when an abutment being lost, bearing in mind that the patient has two temporary teeth which are susceptible to be lost later. Since the placement of implants was not possible in the posterior region without complex procedures such as nerve repositioning and iliac bone graft, three narrow-diameter implants (JD EVOLUTION® S, Italy) were planned to be placed in the interforamina region.

CBCT was done to know the exact bone width and height and the location of the mental foramen. Three dental implants were placed in the anterior mandible between the mental foramina with (3.2 × 10, 3.2 × 8, 3.2 × 8 mm) diameter for the right, middle, and left implants, respectively, and insertion torque 60 N. Cm^{-2} for the right implant and 45 N. Cm^{-2} for the others (Figure 4).

A synthetic bone graft (Cerabone®, Botiss Biomaterials), with a membrane (Jason® Membrane, Botiss Biomaterials),







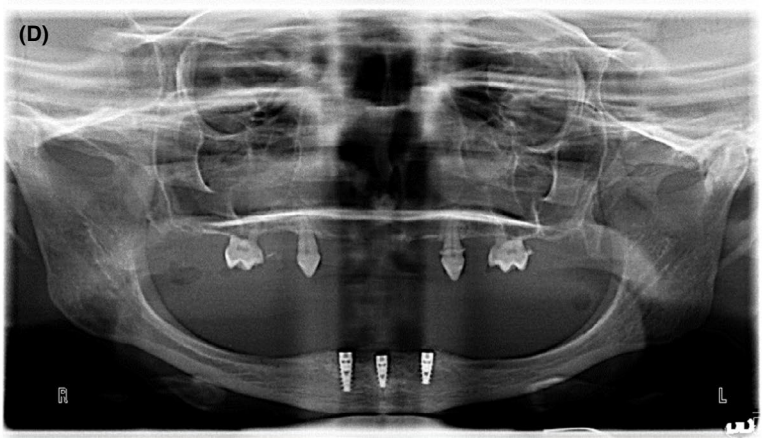
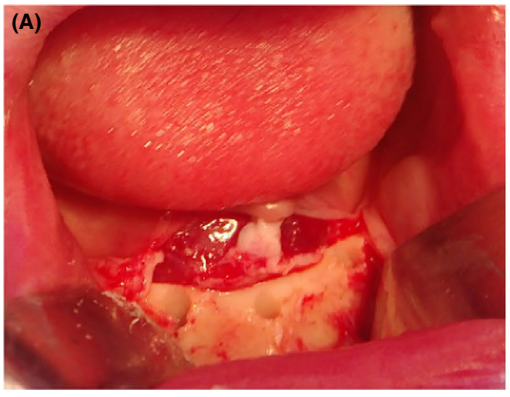
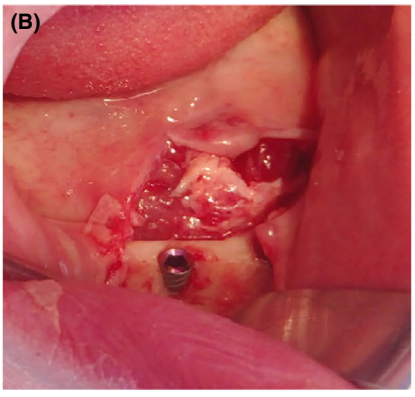


FIGURE 4 (A) Presurgical planning. Note the dense bone near the right mental foramen and the sand clock shape of the bone at the longitudinal section view. (B + C) Insertion torque. (D) After implants insertion





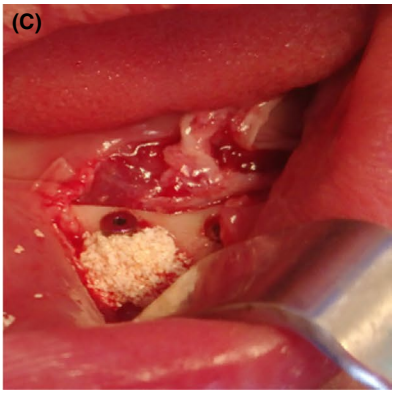


FIGURE 5 (A) Absence of bleeding, (B) implant exposure, and (C) gingival graft

was used to cover the buccal-cervical exposure of the right implant (Figure 5).

The surgical sites were closed with sutures. A standard course of antibiotics and analgesics for the first 48 h after implant surgery was prescribed to reduce swelling and post-surgical discomfort. The patient was also advised to use an antimicrobial mouthwash.

Sutures were removed after 10 days. The patient was recalled after 3 months, for implant second stage surgery, but the right implant failed as the bone in its location was very dense with poor blood supply, which will lead to less chance of adequate healing, so we depended on the two remaining implants.

Healing abutments were placed. Primary impressions were made for fabricating trial denture bases, which will serve as an index for preparing maxillary teeth, also as a guide for the laboratory technician during the telescopic crowns milling process. Minimal preparation was done with the elimination of undercuts, in which we avoided the need for endodontic therapy. Maxillary special tray was made on the master cast, on which primary and secondary crowns were milled.

As the patient has insufficient attached gingiva width in the mandible, a gingival xenograft (Geistlich Bio-Gide®) and autograft were used to increase its width, in a context of a clinical trial held at periodontology department (Figure 6).

Maxillary pick-up impression was made after cementing the primary crowns with zinc polycarboxylate (ADHESOR™ CARBOFINE), during the healing period of the mandibular gingival graft. As the right canine was buccally inclined, we did not make a secondary crown for it, for two reasons: firstly, to avoid modifying its axis during preparation and the subsequent need for endodontic treatment and secondly, to avoid interference of the crown with the arrangement of the acrylic teeth.

The maxillary master cast was obtained (Figure 7).

After 6 weeks, the patient was recalled for making a mandibular primary impression, in order to make a special tray. The final impression was made with Aquasil ultra monophase medium body addition silicone impression material (Dentsply Caulk) instead of using ZOE, which will make the patient's lips swelled because she has decreased secretion of saliva.

The mandibular base was fabricated; then, a definitive interocclusal relationship was obtained.

After the try-in session, the prosthesis was cured and delivered (Figure 8). Ball attachments (Ball abutments H 1.5, H 3.0 JDEvolution® S) were used. Secondary crowns were fixed indirectly.

The patient was educated about the proper methods to achieve acceptable oral hygiene after implants through models and videos. She was also instructed to continue oral hygiene visits every 4–6 months during postoperative years.

3 | DISCUSSION

This is a case report of a hypohidrotic ectodermal dysplasia patient, complaining of a lack of chewing ability and an unpleasant appearance of her jaws. Cephalometric measurements exhibited that EDs tend to have mild mandibular prognathism in the females with a tendency to retruded maxilla and a significant reduction in lower facial height due to hypodontia and poor development of alveolar ridges.[17]

Treatment plan was decided as telescopic overdenture in the maxilla because this type of retainers provides support, protection from dislodgement, and transfers bite forces along the long axis of the abutment teeth.[18,19] Crowns were designed and milled precisely with CAD/CAM technique.

As dental implants placed in ED patients, either infants or adults, present a high survival rate 20-year CSR (clinical survival rate) 84.6%,[18] dental implants were inserted in the anterior part of the mandible, in order to improve the retention of the mandibular overdenture.[20]

It is recommended to insert implants in completely edentulous patients at an early age if the patient has the potential to be compliant, because the long-term use of a conventional denture may be destructive to the bone and soft tissue of the denture bearing area.[1] With taking into account the status of skeletal growth, existing

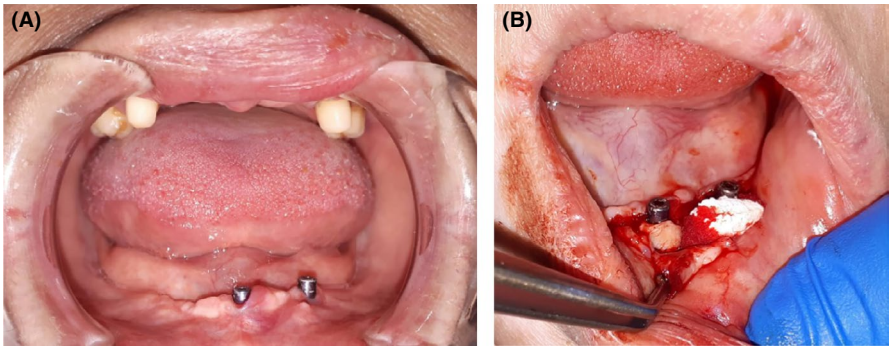


FIGURE 6 (A) Lack of attached gingiva and (B) gingival grafts

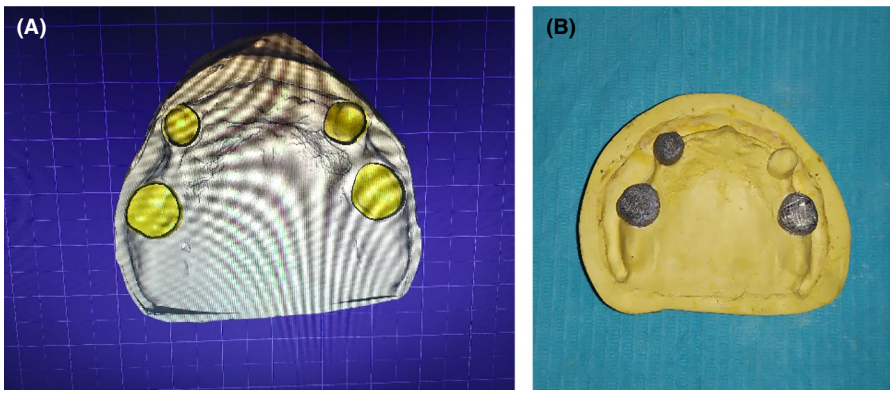


FIGURE 7 (A) CAD-CAM milled crowns, (B) master cast

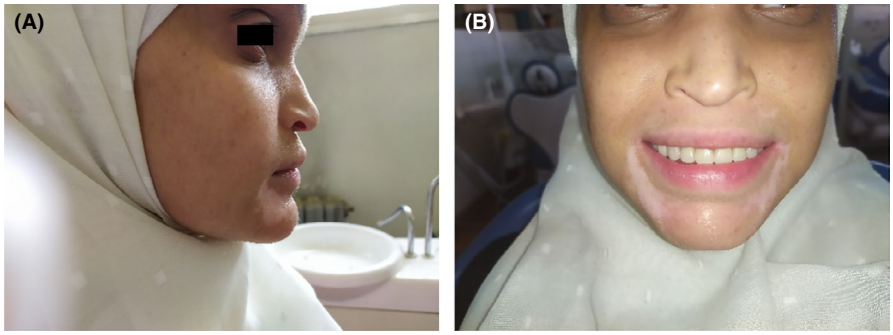


FIGURE 8 (A) Improvement in patient's profile (B) postoperative view

dentition, the severity of hypodontia, and compliance of the patient.[20]

The new denture restored the patient's VDO and improved her appearance, speech, and masticatory function, which was reflected in her psychological well-being, self-confidence and improved her dietary nutrition.

4 | CONCLUSION

EDs are usually being bullied especially at school age because of their unfamiliar appearance, whereas they will struggle with significant psychosocial issues, affecting their self-estimation negatively. In addition, lack of dentition will lead to critical problems in mastication, diet, and growth. All previous problems can be avoided through early diagnosis and appropriate dental treatment, which includes overdentures for children and either fixed or removable prostheses for adults. Also, implants are a good option provided that the patient maintains good oral hygiene and preserves periodical examinations, and it is advantageous to place implants as early as possible for EDs to preserve the volume of the alveolar bone.

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

HA performed the clinical procedures of this case related to the rehabilitation of ectodermal dysplasia patient, wrote the draft for the case report and introduction and discussion Sections, and was responsible for the submission. JS did the implant surgery and critically reviewed the final draft of the manuscript. All authors have critically reviewed and approved the final manuscript and are responsible for the content.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

Data sharing not applicable—no new data generated, or the article describes entirely theoretical research.

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