

INTRODUCTION

Mowat–Wilson syndrome is a rare congenital genetic disorder caused by heterozygous pathogenic variants or deletions of the *ZEB2* gene. It is characterized by distinctive craniofacial features, intellectual disability, and multiple congenital anomalies, most commonly affecting the gastrointestinal, neurological, cardiac, and genitourinary systems. Hirschsprung disease is frequently associated and may serve as an early diagnostic indicator. Clinically, affected individuals often present with broad nasal tips, hypertelorism, large uplifted earlobes, and a prominent chin, along with delayed psychomotor development and seizures. Oral manifestations may include delayed tooth eruption, malocclusion, bruxism, and behavioral challenges, complicating dental management.

CASE REPORT

A 16-year-old male initially presented to the Emergency Department at University Hospitals St. John Medical Center and was subsequently transferred to the Emergency Department at University Hospitals Rainbow Babies & Children's Hospital in May 2025 following a fall from his wheelchair down two stairs that resulted in facial trauma. His past medical history was significant for Mowat–Wilson syndrome, atrial septal defect, seizures, prior cerebral infarction, developmental delay, tracheal stenosis, hemiparesis, undescended testes, and recurrent nephrolithiasis. His surgical history included a tracheoplasty slide procedure and laparoscopic orchiopexy performed in 2020. Current medications included Prilosec, sodium bicarbonate, Pyridium, and Depakene. Reported allergies included cheese, milk, and propofol.

Extraoral examination revealed a 3-cm submental chin laceration (Fig. 2) that had been sutured at St. John's emergency department, along with significant right-sided mandibular and submandibular swelling and limited mouth opening (Fig. 1). Intraoral examination demonstrated a hematoma involving the floor of the mouth and displacement of the right mandible. Tooth #28 was displaced lingually but remained non-mobile, and all observed anterior dentition also presented as non-mobile. Intraoral examination and definitive dental diagnosis were limited by restricted mouth opening, excessive salivary and blood pooling within the oral cavity, and the inability to obtain dental radiographs. Computed tomography (CT) imaging demonstrated a comminuted fracture of the right mandibular body with displacement of fracture fragments by approximately 5 mm in the anteroposterior dimension (Fig. 5) and separation of approximately 11 mm in the transverse dimension (Fig. 6). The fracture extended through the alveolar ridge surrounding unerupted right mandibular premolars and supernumerary teeth, with portions of the lingual alveolar ridge projecting into the floor of the mouth (Fig. 8). A second comminuted fracture was identified involving the left mandibular condyle, with slight inferior displacement of the medial fracture fragment by approximately 1–2 mm (Fig. 7). CT imaging also revealed multiple over-retained primary teeth (#A, B, C, H, J, K, L, and T) (Fig. 3-4), bruxism, and supernumerary teeth in the lower right (Fig. 8, 10) and left quadrant (Fig. 9, 11), findings that the patient's home dentist had made aware to the patient's parents. Management of this case required a multidisciplinary approach involving consultation of both dental and plastic surgery teams. The patient was admitted and underwent reconstructive surgery the following day under the service of plastic surgery to repair the fractures of the right mandibular body and left condyle using internal fixation plates and rubber bands.

CLINICAL AND RADIOGRAPHIC PRESENTATION



Fig. 1

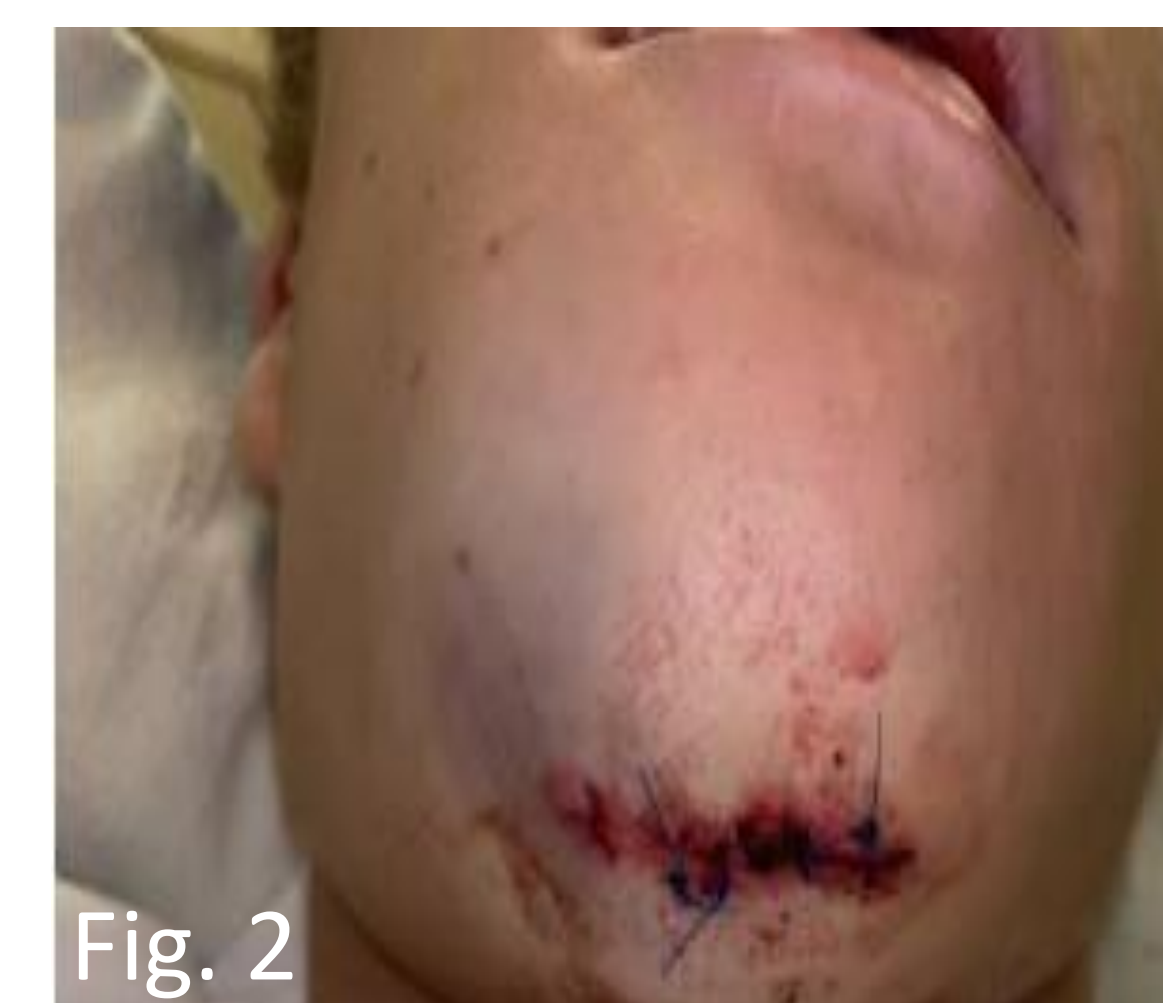


Fig. 2



Fig. 3



Fig. 4



Fig. 5

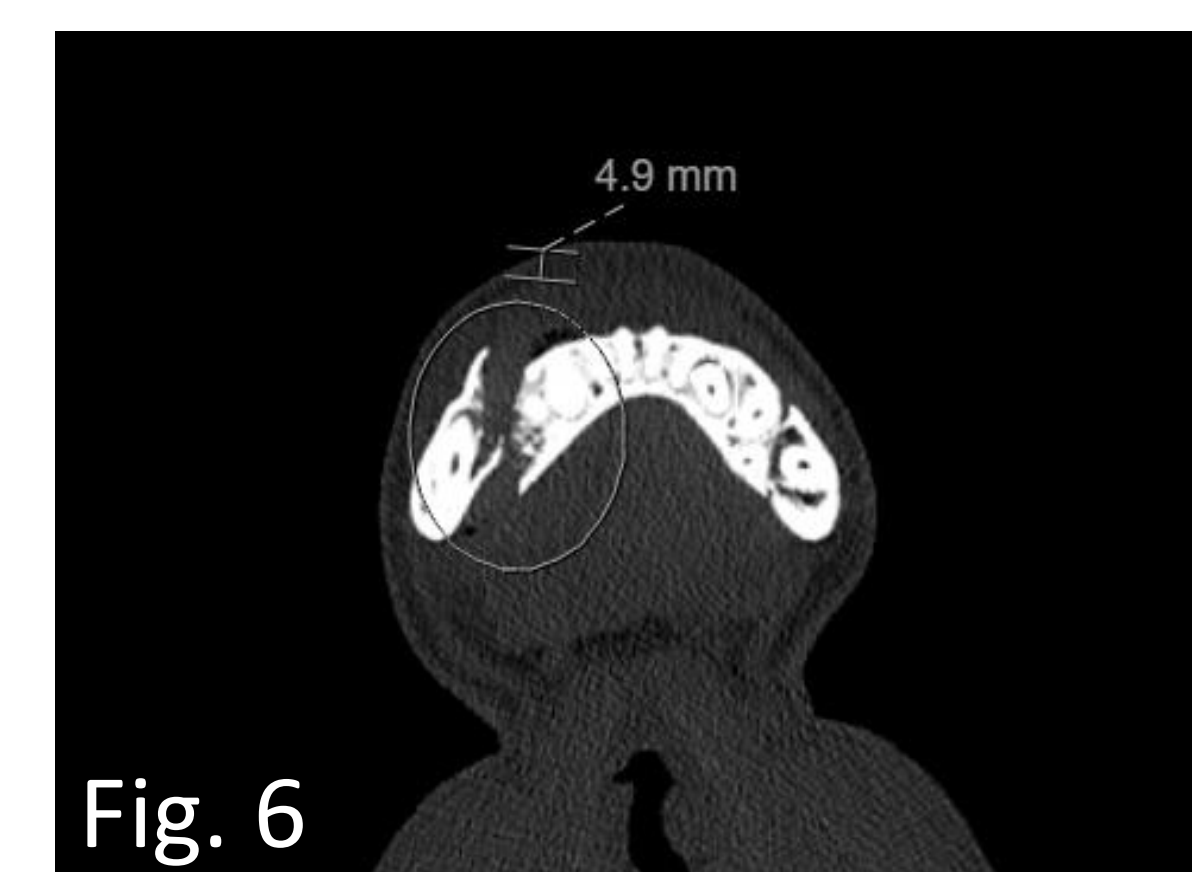


Fig. 6



Fig. 7



Fig. 8

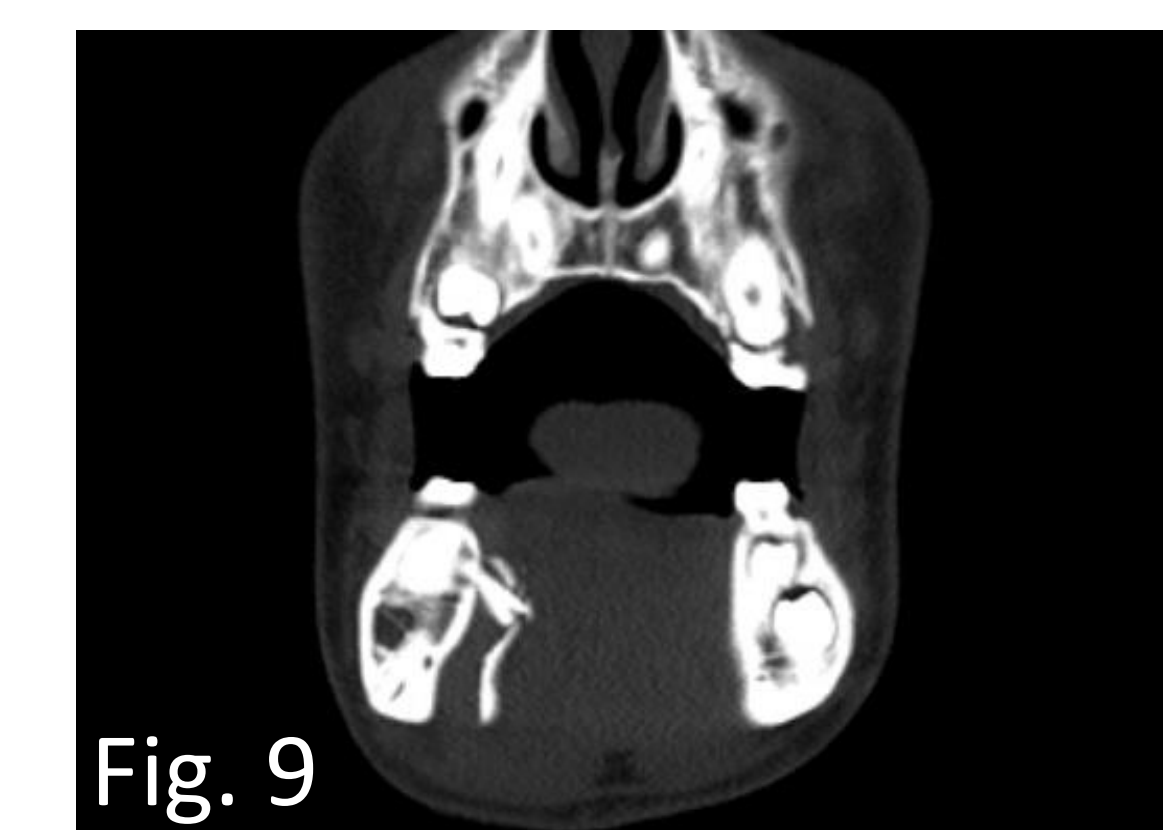


Fig. 9

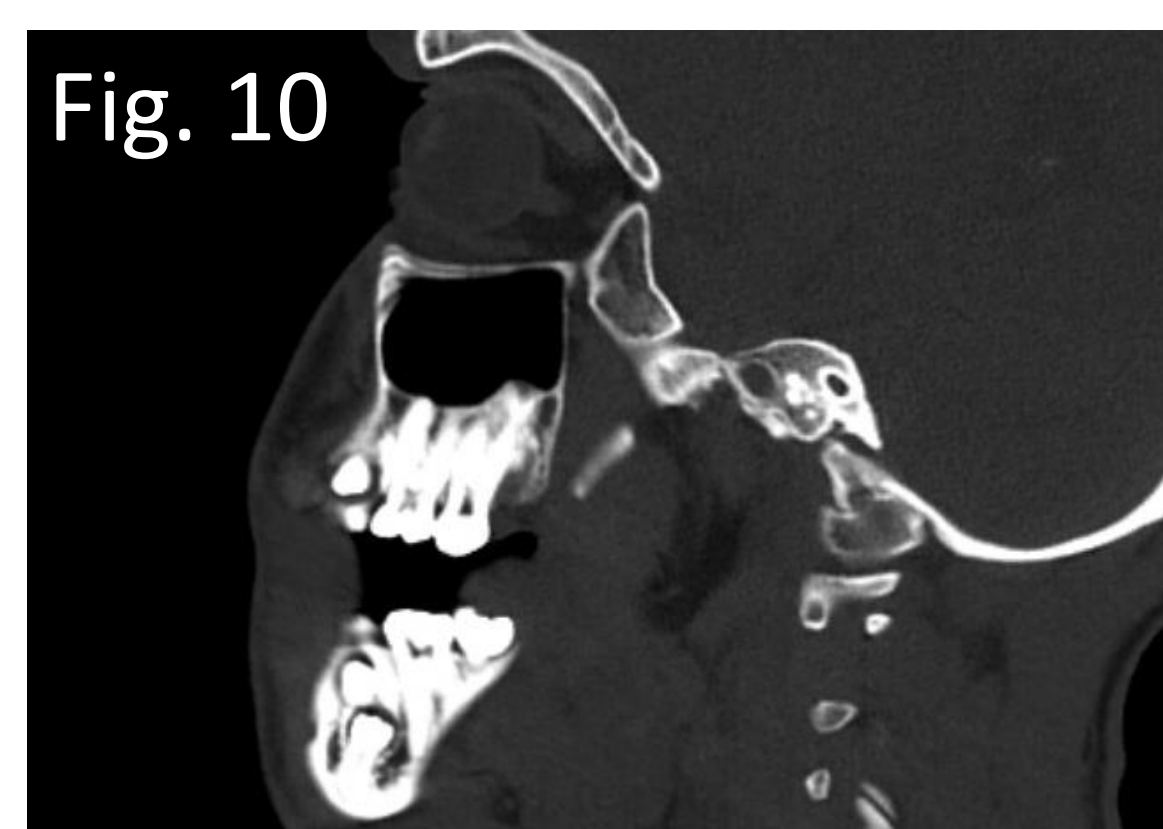


Fig. 10

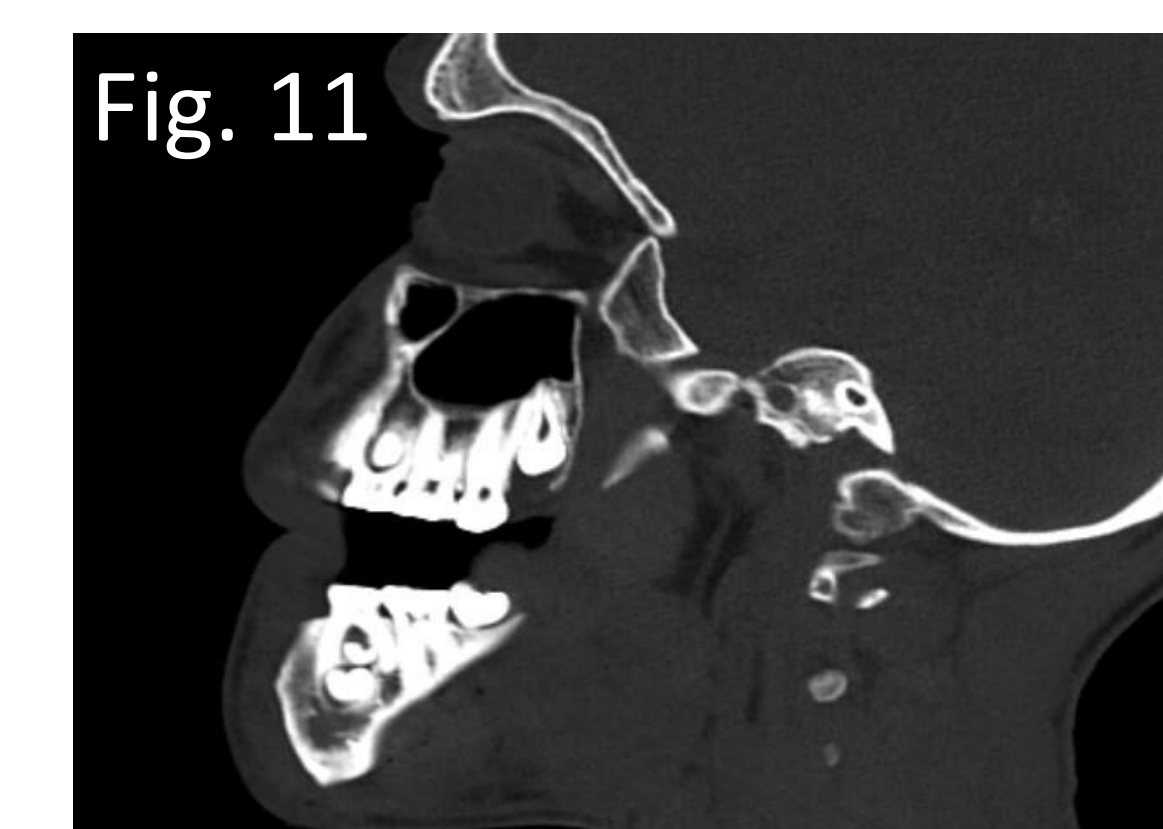


Fig. 11

DENTAL & CRANIOFACIAL FINDINGS

The following dental and craniofacial anomalies were noted in our patient which is typical for a patient with Mowat–Wilson Syndrome:

- Microcephaly
- Delayed tooth eruption
- Bruxism
- Supernumerary teeth

DISCUSSION

Mowat–Wilson syndrome (MWS) is a rare genetic disorder associated with several dental findings, including widely spaced teeth, delayed eruption, malpositioned teeth, and dental crowding. Individuals with MWS may also present with oral health concerns such as bruxism, gingival enlargement, and oral motor dysfunction, which can contribute to feeding difficulties and additional oral health challenges. Dental care for patients with MWS can be complicated by these factors, as well as developmental delay and behavioral considerations, and may require modified behavioral guidance techniques or treatment under sedation or general anesthesia. Preventive dental strategies—including caregiver education, topical fluoride application, and frequent recall visits—are essential to reduce the risk of oral disease and maintain oral health.

The patient described in this case routinely attends dental periodic examinations. The patient's primary dentist has discussed with the parents the plan to continue monitoring the delayed exfoliation of the primary dentition, with treatment under general anesthesia only if clinically indicated. Due to the presence of multiple supernumerary teeth, delayed eruption of permanent teeth, and significant trauma to the area with these unerupted teeth, surgical intervention may be necessary in the future. This could involve removal of the supernumerary teeth as well as surgical exposure of unerupted permanent teeth to facilitate eruption.

Orthodontic treatment may benefit the patient once the permanent dentition has erupted; however, behavioral limitations and the patient's ability to tolerate repeated appointments may impact the feasibility of comprehensive orthodontic care. Early dental involvement and interdisciplinary collaboration with medical providers and dental specialists are essential to optimize treatment planning, improve quality of life, and support long-term oral health outcomes in patients with MWS.

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