

# Jacob's Disease, a rare Disease of the Coronoid Process of the Mandible

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## Abstract:

Jacob's disease is a very unusual pathology with very few cases reported in the literature. In most of the reported cases, this disease mainly affects the left side in young patients. We present a case of a 15-year-old patient with right Jacob's disease and a restriction of the mouth opening. The patient required an intraoral coronoidectomy and subsequent histopathological study. The final result of the pathology reported an osteochondroma of the coronoid process. When presenting a difficulty for mouth opening, researchers must think of different differential diagnoses. Cases of this disease must continue to be reported in order to have significant statistical values throughout the world and to know this pathology in greater depth.

**Keywords:** Jacob's disease, Osteochondroma, Coronoid process.

## Introduction

Jacob's disease is not frequently reported. It was first reported by the doctor Langenbeck in 1853 but it was the French anatomist Oscar Jacob that in 1899 described osteochondroma of the coronoid process forming a pseudoarthrosis between the coronoid process and the zygomatic arch.<sup>1,2</sup>

In published cases, the disease involves males to 63% more often than females, mean age of 30 years with an age range of 5 to 73 years. In published cases, there is a slight tendency for involvement of the left coronoid process.<sup>3</sup>

The pathogenesis of this disease remains uncertain. Clinically, this disease is characterized by a progressive limitation of the mouth opening, generally asymptomatic, and can cause facial asymmetry. A movable lump over the zygomatic arch can be palpated by mandibular movements and the mandibular opening deviates to the affected side in unilateral cases.<sup>4,5</sup>

Examination of images is essential to make a definitive diagnosis. Plain radiographs help identify some features of the injury. Tomography can show the bone lesion in more detail, as well as represent the presence of calcifications, which is why it is considered the gold standard and is of great value for surgical planning.<sup>6</sup> This disease rarely features in the literature. We describe an unusual case of Jacob's disease in a patient who presented with limited oral opening and required surgical intervention intraorally.

## Clinical Report

A 15-year-old male patient consults the Maxillofacial Surgery service because he cannot open his mouth. The patient and relatives deny personal pathological history, deny hormonal alteration or developmental alteration. Clinically the patient had a mouth opening of approximately 10 millimeters (Fig 1). The patient denied the presence of other symptoms, so a panoramic radiograph and a face tomography with three-dimensional reconstruction were indicated.

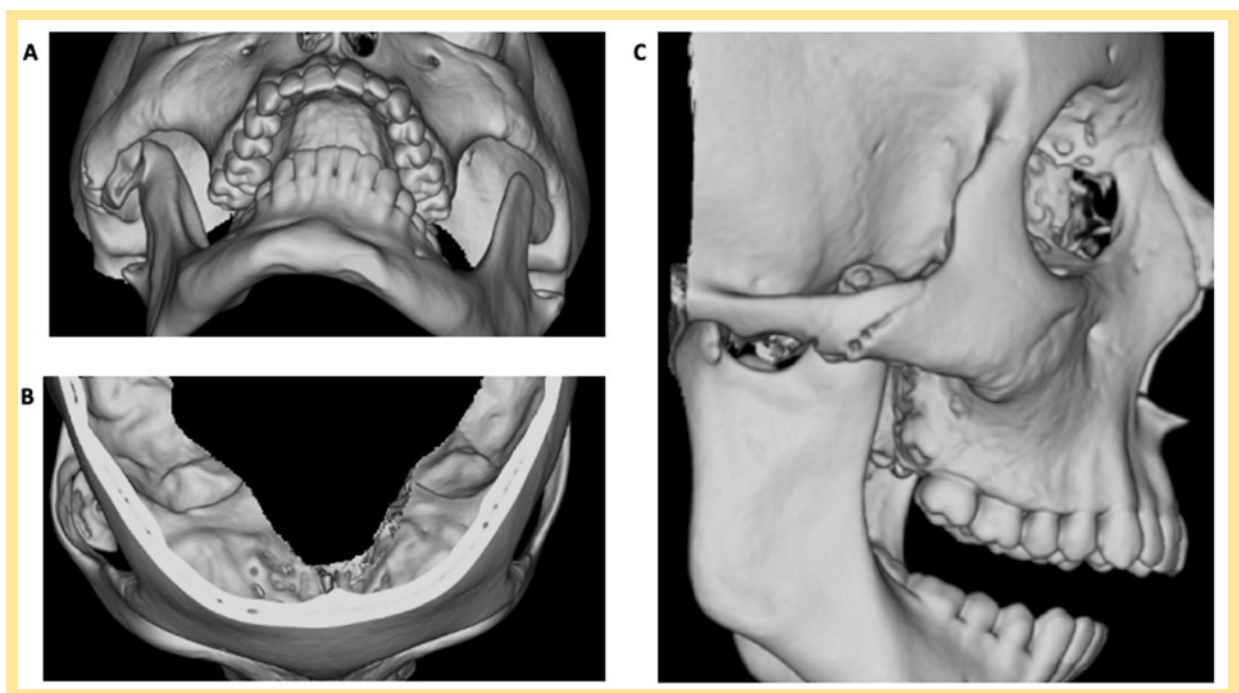


**Fig 1:** Mouth opening of the patient at the moment he enters the medical consultation.

Panoramic radiography (Fig 2) shows an increase in the coronoid process with a cone shape on the right side of the mandible. This anatomical alteration is corroborated when observing the tomography with three-dimensional reconstruction (Fig 3). The tomography shows an increase in growth at the level of the coronoid process of the mandible on the right side. The right coronoid process maintains a heterogeneous amorphous structure and no litigation activity is observed.



**Fig 2:** Panoramic x-ray showing an enlargement of the right coronoid process.



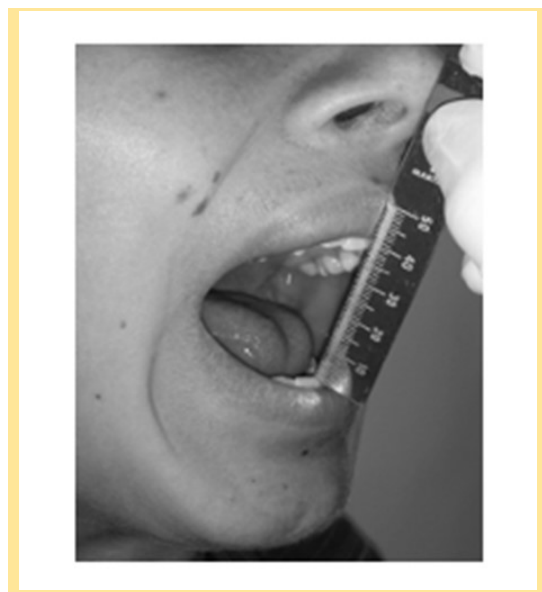
**Fig 3:** Face tomography with three-dimensional reconstruction. A displacement of the zygomatic arch is observed with a pseudo-articulation with the coronoid process.

Initially, when observing the images, a pathological process was considered at the coronoid level and the patient was scheduled for surgery. A coronoidectomy was performed using an intraoral approach under general anesthesia. The coronoid with the lesion was resected and sent to pathology (Fig 4). The surgery was carried out without complications in 2014.



**Fig 4:** Coronoid segment resected intraorally.

The patient recovered his mouth opening to approximately 40 mm (Figure 5). Pathology corroborated the presence of an osteochondroma in the conoid process. The patient has not presented complications until 2020.



**Fig 5:** Final mouth opening of the patient after surgery. Control of the patient at three months.

## Discussion

Jacob's disease is the name given to a pseudo-joint formation between the coronoid process and the zygomatic arch due to an osteochondroma of the coronoid process. Osteochondromas have been described in the mandible, mainly in the mandibular condyle and in the coronoid process. This pathology is known to arise from the metaplastic cartilage generated by the periosteum and it has been reported that excessive stress caused by the temporal muscle could be its cause. This can explain the apparition in the coronoid process of the mandible, although genetic or hormonal causes have also been described.<sup>7-9</sup> The cases of this pathology are few. Of those reported in the literature, some bilateral cases have been described, although most reported cases are unilateral.<sup>3, 10</sup>

This can explain the apperition in the coronoid process of the mandible, although genetic or hormonal causes have also been described.<sup>7-9</sup> The cases of this pathology are few. Of those reported in the literature, some bilateral cases have been described, although most reported cases are unilateral.<sup>3,10</sup> Sreeramaneni<sup>3</sup> carried out an analysis of the literature in search of confirmed Jacob's disease from 1943 to 2010. The researchers found only 51 published cases throughout this near 70-year period. It is possible that a professional throughout her or his career may never see a patient with this pathology.

When professionals have a patient with limited mouth opening and do the diagnostic imaging analysis and identify an increase in the coronoid process of the mandible, they must think of different pathologies. They must first think of hyperplasia of the coronoid process, although if the coronoid has an alteration of the morphology and forms a pseudo-joint with the zygomatic bone, it can be penalized in the presence of an osteochondroma or Jacob's disease. On the other hand, the final diagnosis should always be taken with histopathological confirmation.<sup>1,11</sup>

Coronoidectomy is the definitive treatment for this disease. Although it is preferable to carry out the procedure using an intraoral approach, the approach described in the literature varies. Many cases have been reported as taking an extraoral approach.<sup>12</sup> In addition, the coronoidectomy combined with simultaneous reduction malarplasty could be an alternative method for the treatment of this pathology.<sup>6</sup>

## Conclusion

This pathology is very rare, so every specialist in the head and neck should know it. When a patient presents with difficulty in mouth opening, different differential diagnoses must be considered. Cases of this disease must continue to be reported in order to have significant statistical values throughout the world and to deepen knowledge of this pathology.

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