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# Progressive Limitation in Mouth Opening: Jacob Disease. — Source link

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additional valuable information that is helpful in the diagnostic decision making such as intracranial extension.

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# Progressive Limitation in Mouth Opening: Jacob Disease

**To the Editor:** We would like to discuss with you a rare condition known as Jacob disease. The first description of coronoid process enlargement was by Langenbeck[1] in 1853, but the first report of an osteochondroma of the coronoid process, forming a pseudojoint between the coronoid process and zygomatic arch, was attributed to the French anatomist Jacob[2] in 1899. Currently, only 42 patients of Jacob disease have been reported and confirmed by histologic evidence.[3,4] Additionally, to our knowledge, our patient is the sixth typical patient of mushroom-shaped Jacob disease—only 5 similar patients having been reported.[5–9]

## THE PATIENT

A 59-year-old female presented at our department with a 1-year history of progressive limitation in mouth opening. She reported no pain or other symptoms. No relevant medical history was elicited, and no trauma reported. Physical examination revealed a maximum mouth opening (MMO) of 9 mm with a mandibular deviation to the right side (Fig. 1A). She also presented facial asymmetry with a mild swelling in the right malar region. There was no involvement of the temporomandibular joint on the right or left side. Painless rigid end-feel was also present.

A panoramic radiography revealed abnormal elongation of the right coronoid process overlapping the ipsilateral malar bone and computed tomography, showed a mushroom-shaped coronoid process enlargement and accommodation of the distal coronoid process on the inside of the malar bone (Fig. 1C).

Surgery was performed under general anaesthesia using an emicoronal approach. A joint structure was observed between the enlarged coronoid process and the inner surface of the malar bone with visible cartilage between the 2 surfaces. We performed right coronoidectomy and remodeled the malar bone. In the immediate postoperative period, a 44-mm MMO was obtained.

Two separate bone fragments were sent for histological evaluation. The first consisted of an elongated structure of about 1 cm in length with a round, mushroom-like extremity that showed histologically features of an osteochondroma with a thin cortical envelope, normal trabeculae, and a thick hyaline cartilagineous cap overlying endochondral ossification (Fig. 1E and F). The second fragment removed from the attachment of the first to the mandible showed a nodular bone lesion 0.5 cm in diameter, with features of a compact variant of osteoma with few fibrovascular spaces within compact lamellar bone (Fig. 1G).

From the first postoperative day, the patient underwent joint physiotherapy, passive for the first week and then active for the following 3 months. Six months after surgery, the patient has a 49-mm MMO with no deviation during mouth opening (Fig. 1B and D). The aesthetic results are satisfactory with improvement of facial asymmetry.

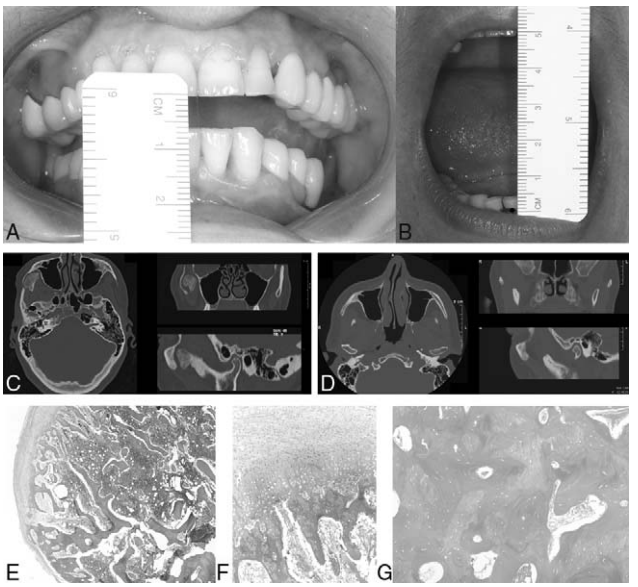


**FIGURE 1.** (A) Preoperative MMO; (B) postoperative MMO; (C) preoperative CT; (D) postoperative CT; (E) extremity of the elongated structure showing a thick cartilagineous cap, the bone trabeculae are regular in size and spacing and the lacunae contain hematopoietic tissue (H&E, original magnification ×4); (F) detail of the cartilagineous cap showing chondrocytes embedded singly in lacunae within the chondroid matrix, with features of regular maturation toward the surface and of endochondral ossification toward its base (H&E, original magnification ×20); (G) detail of the osteoma, showing compact lamellar bone with few fibrovascular spaces (H&E, original magnification ×20). CT, computed tomography; MMO, maximum mouth opening.

## DISCUSSION

Osteochondroma is a cartilagineous lesion. The true nature of the process, whether developmental, reactive or neoplastic, is unknown.[10] It is thought that the proliferating cartilage cells may derive from displaced epiphyseal cartilage cells, from stimulated mesenchymal cells of tendinous insertions, or from pluripotent periosteal cells.[5,10] An unusual feature of the patient herein presented is the presence of a relatively small lesion at the basis of the osteochondroma showing the features of the compact variant of an osteoma. Solitary osteomas of the jaw are rare, mostly occurring on the lingular aspect of the mandibular angle but may also involve the coronoid process or condylar notch.[10] It can therefore be suggested that the osteochondroma in the present patient is reactive in nature, due to the presence of the underlying osteoma.

The aetiology of progressive limitation in oral opening can arise from extra- or intra-articular pathology. The presence of an intra-articular pathology, like chronic temporomandibular joint disk displacement, can lead the clinician to neglect Jacob disease when the 2 conditions coexist in the same patient. Therefore, we recommend a radiographic study: a screening panoramic radiograph can show enlargement of the coronoid processes, and magnetic resonance imaging can rule out internal derangement and joint pathology; computed tomography can confirm the absence of temporomandibular joint ankylosis and will clearly show coronoid enlargement.[11]

Coronoidectomy is the definitive treatment for Jacob disease. According to our experience, advantages such as direct vision, easy and safe osteotomy control, minimal risk of damage to the medial tissue, bloodless operatory field, fast surgery, and brief hospitalization can be obtained using endoscopically assisted intraoral coronoidectomy.[12] When exostosis is bulbous and large, such as in our patient of a mushroom-shaped lesion, reconstruction of a deformed zygoma with a calvarial bone graft is required, or a bilateral condition exists, we recommend a coronal or an emicoronal approach. The latter approach guarantees a good view of the surgical field and results in an acceptable scar behind the hairline; but the risk of damage to the facial nerve remains.[8] Based on the authors' experience, during the osteotomy, the best control and maximum cutting precision can be achieved using the ultrasonic bone-cutting technique.[13]

## CONCLUSION

Additional studies are needed to define aetiology of Jacob disease, but our study represents a supporting evidence to reactive nature of the lesion.

In patient of progressive limitation in mouth opening the coexistence of an intra-articular pathology can lead, even experienced clinician, to neglect Jacob disease, therefore radiographic study is recommended.

For intraoral coronoidectomy endoscopic assistance is advised, but large lesion can justify extraoral approach.

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# Oral Reconstruction Using Medial Sural Artery Perforator Flap in a Patient With Fanconi Anemia

**To the Editor:** Fanconi anemia (FA) is a rare genetic disorder characterized by congenital malformations and aplastic anemia in childhood; followed by hematologic malignancies and solid cancers such as squamous cell carcinomas of the head and neck region