

## A DIGITAL MALFORMATION: CASE REPORT

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**ABSTRACT**

Syndactyly is a very common malformative abnormality of the hand. The diagnosis is most often made at birth. We report an exceptional case of a 33 year old women consulting for the first time for a bilateral complex syndactyly.

**INTRODUCTION**

Syndactyly is a very common malformation of the hand. Diagnosis is most often made at birth or antenatally. Syndactyly may be isolated or part of a malformative syndrome. Imaging is essential to assess associated anatomical damage.

**CASE REPORT**

A 33-year-old female patient, with no previous history, was presented with a congenital malformation of both hands. She presented with a fusion of the 3rd commissures, between the 3rd and 4th fingers bilaterally with a flexion deformity of the latter (Fig. 1). Clinical examination revealed complete fusion of the third and fourth fingers with significant functional limitation, raising suspicion of underlying skeletal anomalies. The rest of the examination did not reveal any associated malformation. The symptomatology dated back to birth. Due to tribal and ethnic beliefs, the patient had not previously sought medical attention. A standard X-ray and CT scan of both hands were performed (Fig. 2), showing bilateral complex syndactyly.

**DISCUSSION**

Syndactyly is the partial or complete fusion of adjacent fingers. This malformation may involve the skin covering, bone, tendons and vascular and neural elements to varying degrees. It may be isolated or part of a malformative syndrome.<sup>[1]</sup>

The isolated form is most often secondary to autosomal dominant familial transmission with variable expression. Our patient's case was isolated in her family, no similar cases were found.

Pathophysiologically, syndactyly is secondary to a defect in the separation of the digital rays as a result of an

abnormality in the programmed apoptosis process that is designed to separate the digits during the sixth week of embryonic development.<sup>[2,3]</sup>

The diagnosis is clinical and is made at birth (antenatal diagnosis being difficult, especially for isolated forms). The examination must be bilateral and comparative, involving the upper and lower limbs. The syndactyly must be analyzed morphologically and the impact on mobility must be assessed.<sup>[2]</sup>

Complex syndactyly is characterized by skeletal fusion which may be distal or proximal, whereas simple syndactyly involves only the skin covering.

Imaging is then essential to analyze the bone connections. Standard X-rays or CT scans can be used to describe the fusion points. The fusion may be total or partial, affecting the proximal or distal phalanges or both.

In our patient, on the right side, the fusion was proximal, involving only the first phalanges, in a partial manner, whereas on the left side, the fusion was disto-proximal, with a complete reunion of the proximal phalanges and the existence of fusion bridges between the distal phalanges.

Ultrasound coupled with color Doppler is useful for analyzing tendon structures and vasculo-nervous pedicles before undertaking any surgical repair.<sup>[2]</sup> Indeed, tendon abnormalities are common in complex syndactyly with abnormal insertions, supernumerary or absent tendons. The vascular anatomy is often altered, such as an abnormal division of the common digital artery, a single artery, or even an absence of a vessel in the affected commissure.<sup>[4]</sup>

The management of syndactyly is surgical. The aim of surgery is both functional and aesthetic, and consists of recreating a harmoniously shaped commissure while avoiding retractile bands and residual deformities.<sup>[5]</sup> The ideal age for surgery is around 1 year.<sup>[6]</sup> Syndactylies of the first and fourth commissures, joining fingers of different lengths, result in lateral deviation of these fingers. They should be released early, as early as the third or fourth month, to avoid these deformities. Complex syndactyly should be operated on late at the age of 3 years.<sup>[7]</sup>

Resurfacing of the synostosis separation zone involves a number of techniques ranging from composite grafting combining skin and subcutaneous cellular tissue to complex flap techniques (e.g. Buck-Gramck pulpal plasty).<sup>[8]</sup> Simple forms have a good prognosis. In complex forms, the prognosis depends on the associated skeletal, tendon, muscle and joint anomalies. In our patient, age, joint stiffness and bony abnormalities are poor prognostic factors that may impair active mobility even if the fingers were separated.



**Figure 1: Bilateral syndactyly. The right hand shows syndactyly of the 3rd commissure, between the 3rd and 4th fingers, in irreducible flexion, the skin separation is continuous from the base to the digital tip. On the left side, syndactyly also involves the 3rd commissure with a distal unguinal overlap. The skin separation is not individualised.**



**Figure 2 (a)**



**Figure 2 (b)**

**Figure 2: Standard X-ray (a) and three-dimensional CT reconstruction (b) of the hands, showing partial fusion of the first phalanges of the 3rd and 4th fingers on the right and complete fusion of the first phalanges of the 3rd and 4th fingers on the left. Note the presence of distal fusion bridges in the distal phalanges on the left side.**

#### CONCLUSION

The diagnosis of syndactyly is most often made at birth. The surgical planning of the procedure depends on the bony, tendino-ligamentary and vascular parameters associated with the syndactyly, hence the essential role of imaging.

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