







## Pathophysiology

Saint-Hilaire in 1836 and Darwin in 1871 advanced the concept of development of the human race from primitive animals; thus, they also considered the supernumerary nipple as an atavistic structure deriving from the milk line of mammals. Similarly, ectopic supernumerary nipple found on the vulva may express an atavistic structure because the breasts of dolphins and whales are in that location, or ectopic supernumerary nipple on the back, the scapula, and the shoulder<sup>30,31,32</sup> is reminiscent of the nutria and hutia (rodents) with a similar location of the breasts.

Between the fourth and fifth weeks of embryogenesis, an ectodermal thickening forms symmetrically along the ventral lateral sides of the embryo. This epidermal ridge extends from the axillary region to the inner side of the thigh to form the embryogenic milk (or mammary) line. During the second and third embryogenic months, the glandular elements of the breasts are formed near the fourth and fifth ribs, with regression of the rest of the thickened ectodermal streaks. In the case of failure of a complete regression, some foci may remain to result in a supernumerary nipple. This can develop into a supernumerary complete breast (polymastia) or into any other supernumerary nipple variant according to the Kajava classification.

## History

Usually, the supernumerary nipple remains undetected or asymptomatic. Occasionally, the

supernumerary nipple is noticed only when hormonal changes during adolescence, menstruation, or pregnancy cause increased pigmentation, fluctuating swelling, tenderness, or even lactation.

## Physical

The supernumerary nipple is often overlooked at the first examination of the neonate. It appears as a small pigmented or pearl-colored mark or as a concave or umbilicated spot

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In 75% of patients, it measures no more than 30% of the diameter of the normal nipple (at times no more than 0.2-0.3 cm in diameter). In the other 25% of patients, it is of medium size, as large as 50% of the normal size of the nipple. Rarely, a supernumerary nipple is as large as a normal nipple.<sup>28</sup> It can be mistaken for many other small lesions, most of them hardly noticeable. Note the image below.

Most supernumerary nipples are single, and, when 2 or more (as many as 8) supernumerary nipples are present, they are distributed bilaterally or unilaterally, symmetrically or not. Most supernumerary nipples are located below the regular nipple, while approximately 13% appear above it along the milk line.<sup>20</sup>

When examining adolescent girls (note the image below), the normally developed breast may hide the supernumerary nipple. A number of studies have indicated a preponderance of supernumerary nipples on the right side.<sup>25</sup>

For easier detection of the supernumerary nipple, a wet gauze pad is passed along the

mammary line (milk line) from the axillary region to the upper part of the thigh on each side. This technique is particularly helpful in the dry and desquamating skin of full-term and postterm infants.<sup>20</sup> When the suggested lesion is concave, folding it between fingers shows a typical wrinkling.

A dermoscopic examination of supernumerary nipple shows a pattern similar to dermatofibroma, with both showing central, white, scarlike areas and a peripheral fine-pigment network. Supernumerary nipple also has a cleftlike appearance in the central area, thus allowing differentiation from dermatofibroma.<sup>34</sup>

Approximately 5% of supernumerary nipples are ectopic, located outside of the milk line, such as on the back,<sup>32</sup> the shoulder (note the image below),<sup>35</sup> the limbs, the neck, the face, and the vulva and perineum.

## **Causes**

Familial cases of supernumerary nipple were recorded as parent-child transmission, including 1 report of a family who had supernumerary nipples in 4 successive generations; therefore, autosomal dominant with incomplete expressivity is the accepted transmission of inheritance

## **Histologic Findings**

The histologic features of a supernumerary nipple are identical to that of the regular nipple, including hyperpigmentation, slight hyperkeratosis with epidermal thickening, pilosebaceous structure of Montgomery areolar tubercles, smooth muscle bundles typical of the areola, and possible mammary glands and intradermal straight ducts.<sup>37</sup> A significant increase in the number of clear cells of Toker has been found in supernumerary nipple tissue, indicating supernumerary nipple may be a precursor of extramammary Paget disease

## **Surgical Care**

A protruding (or erectile) supernumerary nipple that causes the patient embarrassment can be easily removed surgically, if desired. Removal using liquid nitrogen cryotherapy has been described.<sup>39</sup> The removal of polymastia or a complete ectopic supernumerary nipple (with breast) is more involved but is indicated in women at high risk of developing breast cancer.

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To avoid an unsightly scar after the removal of a complete ectopic supernumerary nipple, the tumescent liposuction technique has been suggested