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Supernumerary Nipples: An Overview

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Supernumerary nipples (SNs) are relatively common, minor congenital anomalies. SNs usually arise within the embryonic milk lines, but they have been known to form elsewhere. The prevalence of SNs varies, depending on the population. SNs are normally benign entities, but they are susceptible to hormonal changes and disease processes and may signify internal disease.

Supernumerary nipples (SNs) are relatively common, minor congenital anomalies. SNs usually arise within the embryonic milk lines, but they have been known to form in other locations, such as the vulva, neck, back, and thigh.[1-5] During the fourth week of embryogenesis, thickened strips of ectoderm known as mammary ridges or lines (milk lines) appear, extending from the axilla to the groin. Mammary buds start to develop as solid, epidermal downgrowths from the mammary ridges during the sixth week of development. Normally the mammary ridges only persist in the pectoral region where breasts develop, but when they fail to regress, an SN is formed.

When an SN is found outside the milk lines, it may represent the reversion of certain characteristics to a more primitive or ancestral state, as described by the Geoffrey Saint-Hilaire–Darwin theory of atavism.[6,7] An example of an atavism is the formation of SN in the vulva, as this is where dolphins and whales have their breast tissue.[8] Other theories have been proposed. One theory postulates that SN outside the milk lines are due to the displacement of embryologic mammary crests. Another theory is that SN are modified apocrine sweat glands.[8] The classification of SN that was described by Kajava[9] in 1915 is still in use today (Table).

SNs are usually solitary, but as many as 8 SNs have been described in one patient.[10] When there are multiple SNs, they can form unilaterally or bilaterally. They usually are found inferior to the normally located nipples but can be found above them. SN prevalence varies greatly, with a range of 0.22% to 6% of the population.[11,12] The prevalence varies according to ethnicity, sex, geographic region, and method used to determine the presence of SN. Specifically, 1.63% of black American neonates,[13] 0.22% of white Europeans,[11] 0.6% of white American neonates,[14] 4.7% of Arab children,[15] 2.5% of Israeli children,[16] 5% of Japanese women,[17] and 1.6% of Japanese men have SNs.[17] SNs are usually sporadic, but about 6% of reported SNs have been familial cases, which are believed to follow an autosomal-dominant pattern with incomplete penetrance.[18,19]

Normally, SNs pose no medical threat and do not require treatment; however, they are subject to the same hormonal changes and disease processes that affect normal breast tissue. If there is glandular tissue, SNs can enlarge during puberty, swell and become tender premenstrually, and lactate.[4] They are known to develop fibroadenomas,[20] adenomas,[21] cysts,[22,12] abscesses,[3] and mastitis,[3] as well as breast carcinoma.[1,12] They also have been associated with a number of medical conditions. A number of studies have identified a relationship between kidney and urinary tract malformations and SN.[11,23-26] The frequency of urinary tract and renal defects in the general population is 1% to 2%.[18] In patients with SNs evaluated with ultrasound, the frequency of kidney and urinary tract anomalies is estimated at 14.5%, with a stronger association in males than females.[26,27] This correlation is even more pronounced with familial SNs. The frequency of kidney and urinary tract defects

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Supernumerary Nipple Classification[9]

Classification by Kajava	Description
1	Complete SN: nipple, areola, and glandular breast tissue; known as polymastia
2	SN: nipple and glandular tissue without areola
3	SN: areola and glandular tissue without nipple
4	Aberrant glandular tissue only
5	SN: nipple, areola, and pseudomamma, which is fat tissue that replaces the glandular tissue
6	SN: nipple only, which is known as polythelia, is the most common type
7	SN: areola only, which is known as polythelia areolaris
8	Patch of hair only, which is known as polythelia pilosa

Data are from Kajava Y.[9]

in patients with familial SNs evaluated with ultrasound has been found to be approximately 30%.[26] This association between kidney and urinary tract malformations and SNs has not been found in black Americans.[13,14,28] Other medical conditions sporadically associated with SNs include vertebral anomalies,[29] cardiac arrythmias,[30] hypertension, peptic ulcer disease,[7] migraine,[31] neurosis,[32] gonadal hypoplasia,[33] pyloric stenosis, epilepsy,[11] intracranial aneurysm, coronal suture synostosis,[14] atrial septal defect, double gallbladder, malformation of the hand, absence of a foot, accessory spleen, hydrocephalus,[34] and testicular cancer.[35] SNs also have been associated with Turner and Fanconi syndromes,[24,36] among others. It has yet to be determined whether these nonrenal associations occur more frequently in people with SNs than in the general population, but it is likely that they were chance findings.

Usually, treatment of an SN is unnecessary, but if the patient is unhappy cosmetically or if the SN causes discomfort from lactation or tenderness, it can be removed surgically. If the SN has undergone a malignant change, then a more aggressive treatment is necessary; the SN should be excised with a wide margin, and the patient should receive appropriate follow-up treatment.[8] Some physicians also have recommended that patients with SN should receive a renal ultrasound, especially those with familial SN, because of the association between SN and renal anomalies, though this is controversial.[25,26,37]

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