

Tardive expansion congenital hemangioma: A niche within NICH?



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Just when I thought I had grasped the concept of congenital hemangiomas, new data raises novel questions.

Compared with infantile hemangiomas, which grow rapidly after birth and then involute, congenital hemangiomas arise in utero. The subtypes of congenital hemangiomas include rapidly involuting congenital hemangioma (RICH), noninvoluting congenital hemangioma (NICH), and partially involuting congenital hemangiomas (PICH). RICH involutes completely within the first 6 to 14 months of life, NICH grows proportionally with the child without regression, and PICH is the result of the evolution from RICH to persistent NICH-like lesions.¹

Cossio et al² retrospectively analyzed 80 cases of NICH. Nine presented with atypical postnatal growth after a stable period between the ages of 2 and 10 years. Five patients showed new red papules on the surface of the lesion, 2 reported bleeding from the papules, and one developed a pyogenic granuloma. All Doppler ultrasounds and/or MRIs were compatible with a diagnosis of NICH, and a confirmatory biopsy was performed in 4 cases. Two patients received endovascular embolization, and one required further surgery for treatment. The authors concluded that NICH may exhibit significant postnatal growth over time, requiring closer follow-up for longer periods.

Knöpfel et al³ described 2 cases of NICH that demonstrated subtle postnatal growth over time: a 6-week-old girl with a right arm NICH which grew with increasing telangiectasia and a full-term 4-week-old boy whose NICH became elevated and darkened.

In this issue of the *Journal of the American Academy of Dermatology*, Hua et al⁴ reported a

Abbreviations used:

NICH:	noninvoluting congenital hemangioma
PICH:	partially involuting congenital hemangiomas
RICH:	rapidly involuting congenital hemangioma
TECH:	tardive expansion congenital hemangiomas

series of 11 unusual congenital hemangiomas initially presenting as NICH with subsequent tardive expansion of the lesion, naming these lesions tardive expansion congenital hemangiomas (TECH). The infants included 9 boys and 2 girls with lesions of the head and neck (n = 10) and abdominal wall (n = 1). Spontaneous expansion began at the age of 12 to 61 months. Uniform parenchymal and fast-flow vessels were confirmed by imaging. Histologic features included lobules of GLUT-1 negative vessels with focal hobnailed endothelium and stellate centrilobular draining channels. The authors speculate that TECH represents endothelial proliferation in response to boosted hemodynamics, resulting in an imbalance between proliferation and involution. Further scrutiny is required to elucidate how this relates to mosaic missense mutations that alter glutamine at amino acid 209 in GNAQ or GNA11, identified in NICHs and RICHs at variant allele frequencies up to 33%.

Although most NICHs do not require treatment, it has been suggested that surgery should be considered early to achieve the best cosmetic and functional results during adult life.⁵ A multidisciplinary consultative approach is recommended before embarking on other therapies (embolization,

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sclerotherapy, and laser) until adequate evidence for the use of these modalities becomes available.

The observation that a subset of NICH may continue to grow postnatally is important for several reasons. Practically, parents must be informed of this potential to avoid undue angst. Theoretically, understanding this phenomenon could yield potential therapeutic maneuvers not only for NICH, but also other lesions where mutations of GNAQ and GNA11 are influential, such as in phakomatosis pigmento-vascularis, extensive dermal melanocytosis, and uveal melanoma. This niche within the NICH warrants clinical recognition and further research.

Conflicts of interest

None disclosed.

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