Hypohidrotic Ectodermal Dysplasia and Breastfeeding

A Resource for Health Care Providers

Hypohidrotic ectodermal dysplasia (HED) is a genetic condition caused by mutations in the EDA gene. The most common form of the condition is x-linked, but autosomal recessive and autosomal dominant forms of the condition exist as well. The most common feature of HED is the inability to sweat (hypohidrosis), but other structures derived from embryonic ectoderm can also be affected including (but not limited to) the hair, nails, and teeth.

Females affected with HED or are carriers of x-linked hypohidrotic ectodermal dysplasia (XLHED) can present with some features such as sparse scalp hair, varying severity of hypohidrosis, or missing teeth.

Other symptoms of HED can include a lack of or supernumerary nipples. The various presentations of atypical breast development in female carriers of XLHED and how it may affect their ability to breastfeed are summarized below.[1]

Sumary of Key Findings From Wahlbuhl-Becker et al. (2017)

- Significant difference in breast size
- Unilateral or bilateral amastia
- Abnormal nipple development including:
    - Abnormally flat nipples
    - Inverted nipples
    - Supernumerary nipples
    - Reduced or absent glands of Montgomery on the areola

Abnormal breast development has been associated with difficulties with breastfeeding, with some women reporting an inability to breastfeed at all.

Some of the factors that women reported affected ability to breastfeed were:

- Nipples were too flat
- Not enough milk

Key Points for Providers and Lactation Specialists

- Women who are either female carriers for XLHED or affected by HED may experience difficulty with breastfeeding.
- Challenges in breastfeeding may be due to abnormal development of breast tissue and nipples.
- Women who do not have visibly dysmorphic nipple structures may have fewer or absent glands of Montgomery that may be responsible for difficulties in breastfeeding.
- Lactation counseling may be recommended and supportive efforts such as special salves or nipple shields may help in facilitating breastfeeding in women affected by HED.

Women affected by HED may have more problems with breastfeeding than healthy mothers and this should be taken into consideration during lactation counseling. Women should be counseled to ease some of the psychological pressure that comes with challenges with breastfeeding.

References:

1. Wahlbuhl-Becker M, Faschingbauer F, Beckmann MW, Schneider H. Hypohidrotic Ectodermal Dysplasia: Breastfeeding Complications Due to Impaired Breast Development.

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T 618-566-2020 E INFO@NFED.ORG W WWW.NFED.ORG