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REVIEW

Bilateral congenital amazia: A case report and systematic review of the literature

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Summary *Background:* Congenital breast anomalies present challenging management decisions to the plastic surgeon. One must consider the optimal age of reconstruction as well as the ideal surgical technique. Amazia, a very rare condition characterised by a complete lack of breast tissue in the presence of a nipple areolar complex (NAC), is one such congenital breast anomaly.
Methods: A comprehensive systematic review of the literature was performed to examine the various approaches to reconstruction of congenital breast anomalies. From this review, the data compiled included patient demographics and operative details, including type of reconstruction, treatment of the contralateral breast and treatment of the NAC. A case of bilateral amazia is also reported.
Results: Of 178 articles, 13 ultimately met the inclusion criteria and 54 individual patient reconstructions were identified from these papers. At the time of reconstruction, the patients were in the range of 13—54 years, with an average age of 27.6 years. Prosthetic and autologous reconstructions were equally represented (19 patients each, 35.2%; Table 2). Autologous reconstruction with prosthesis was slightly less common (15 patients, 27.8%). One patient was reconstructed using autologous lipo-augmentation only.

Of the 36 cases in which the approach to the NAC was addressed, most (66.7%) were not reconstructed.
Conclusions: Amazia is a very rare congenital anomaly of the breast. This systematic review of the literature highlights the need for better reporting and examination of this type of data to allow for future study and to better advise on decision making regarding the timing of reconstruction, surgical technique and the approach to the NAC.

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As a whole, congenital breast anomalies including hypoplasia, amazia and amastia are common in the paediatric population.[1,2] Despite the relative frequency with which these conditions are encountered in clinical practice, the literature is lacking in quantitative reviews of how to manage these patients. Therefore, a systematic review of the existing literature was conducted on congenital breast anomalies allowing an analysis that provides data summarising the age of reconstruction, surgical technique and management of the nipple areolar complex (NAC). Additionally, we present an unusual case of a patient who presented with congenital amazia.

Case presentation

A 13-year-old female presented to the paediatric plastic surgery clinic with bilateral, congenital absence of the breasts and low-set NACs. Normal pubertal development was noted with Tanner stage V pubic hair and with onset of menses at 11 years of age. At birth, she was found to have right foot postaxial polydactyly, bilateral syndactyly of the second and third toes, left ptosis and amblyopia, frontal upsweep of the hairline and low-set, posteriorly rotated ears. Chromosome analysis, renal ultrasound and skeletal survey were all normal, and family history was unremarkable. The patient was seen by Genetics and a normal chromosomal analysis was confirmed; additionally, a syndrome was not identified. Of note, the patient complained of low self-esteem and social isolation due to the complete absence of breasts. She was diagnosed with amazia.

Physical examination revealed no palpable breast tissue. The patient's NACs were positioned on the chest at the midhumeral point, which many use as a reference point for normal nipple position. However, in this case, their position relative to other landmarks on the thorax made them appear to be sitting too far inferiorly. The NACs were sitting at the ideal position for the new inframammary fold (Figure 1). The distance from the midsternal notch to the NAC was 23 cm on the right and 22 cm on the left, and the proposed breast pocket diameter measured 11—12 cm. Other findings included an accessory nipple on the left abdomen as well as axillary fullness that felt like fatty tissue. Hormone levels were normal and a breast ultrasound revealed no hormone-sensitive glandular tissue. Wrist radiographs revealed fused growth plates with a bone age of 15 years.

The patient was counselled that given her young age and the fact that she was likely still growing, it would be prudent to wait to do the reconstruction. She was suffering from significant psychological disturbance secondary to her deformity. Socially she was isolated and would not participate in age-appropriate activities with her peers. After seeing the patient back several times over a 1-year period, the decision was finally made to proceed with reconstruction. Surgical options were discussed. This patient wanted a small- to moderate-sized breast mound, and thus a postoperatively adjustable implant was selected for her reconstruction.

Surgical procedure

The patient was taken to the operating room for placement of bilateral, subpectoral, dual-plane, postoperatively adjustable saline implants. These were placed via periareolar incisions with a lateral extension. The dissection was extended down to the pectoralis muscle in a subcutaneous plane. On the right side the subcutaneous dissection was done for a slightly greater distance superiorly prior to division of the pectoralis muscle as the NAC was sitting slightly more inferiorly on this side. The pectoralis was released bilaterally from the 3 o'clock to the 9 o'clock position and the pocket was extended superiorly such that the implant would sit in a dual plane once inflated. The final pockets measured approximately 11—12 cm in diameter.

Postoperatively adjustable saline implants (Mentor Smooth Round Spectrum implants) measuring 10.8 cm in diameter were placed. The projection was 3.5 cm and the maximum fill volume between 225 and 270 ml. A remote port was placed in a lateral pocket near the midaxillary line and attached to the expander in a standard fashion.

In-office expansion was initiated 2 weeks postoperatively and terminated at a fill volume of 270 cc. The patient tolerated these expansions well and her implants remained symmetrically positioned on the chest wall (Figure 2).

Approximately 1 year after the placement of the implants, the patient returned for repositioning of the NACs. The patient was taken to the operating room for bilateral free nipple grafts. The donor sites were closed primarily. The grafts were secured in place with a Reston foam bolster such that the nipples were visible. The patient was admitted postoperatively and had some superficial slough of the areola and partial loss of the nipple itself. However, she did heal with an aesthetically acceptable result (Figure 3).

Systematic review

A systematic review of the literature was performed using the PubMed for MEDLINE articles. Search strategies

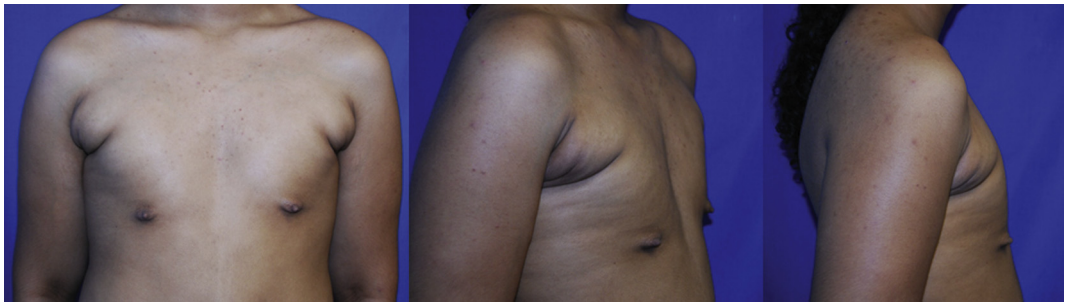

Figure 1 Patient before reconstruction.