Reconstruction of Congenital Arhinia with Stereolithography

Kelly Spiller; Kaitlynne Pak; Lucas Harrison; Spencer Anderson, MD; Steven Schmidt, MD; Salim Mancho, DO Department of Orthopedic & Plastic Surgery

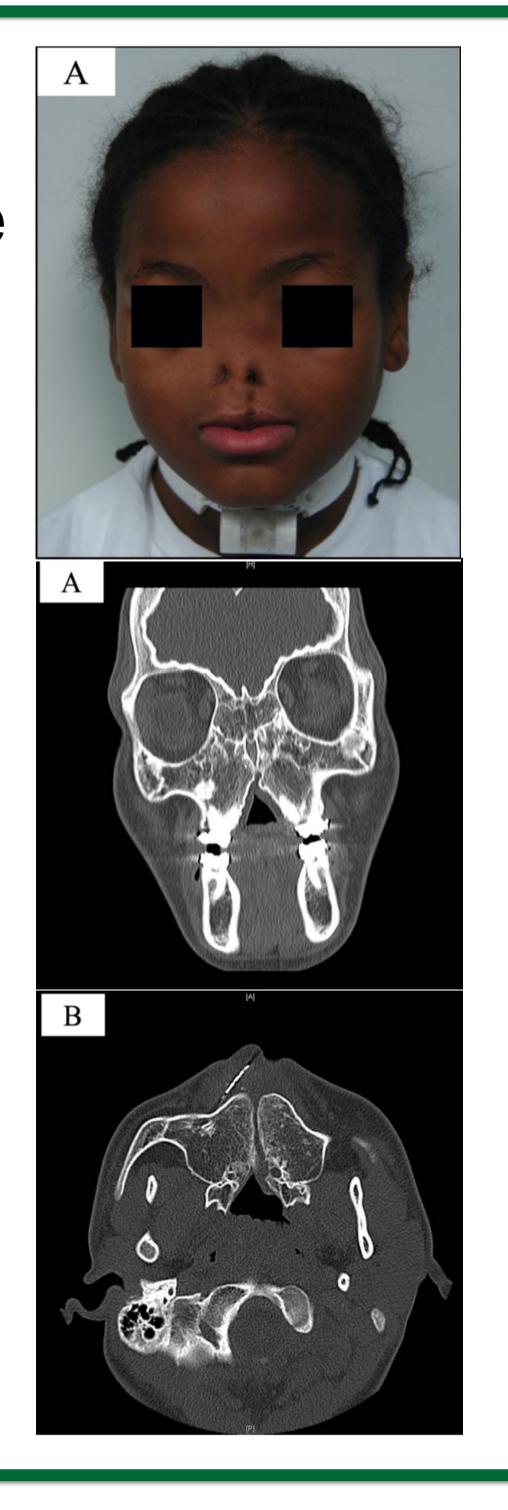


Introduction

- Complete arhinia- rare embryologic disorder, unknown etiology
- <50 reported cases
- Often associated with congenital abnormalities of eyes, ears, palate
- Challenges with breathing, feeding early in life
- Paramedian forehead flap with cartilaginous + bony grafts most frequently reported method of external nasal reconstruction
- We present a unique alternative method of reconstruction using stereolithographic modeling with combined conchal bowl and osteocartilaginous grafts

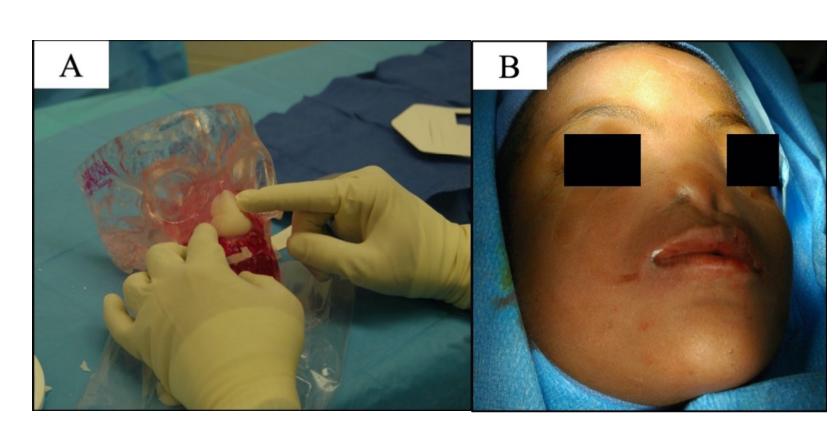
Case Presentation

- 11 yo M presenting with complete congenital arrhinia, hypoplastic midface, and choanal atresia
- S/p L cleft lip repair, tracheostomy, and red rubber catheter placement
- Family refused forehead flap due to significant scar burden
- Goals:
 - Creation functional nasopharyngeal airway
 - Improved aesthetics

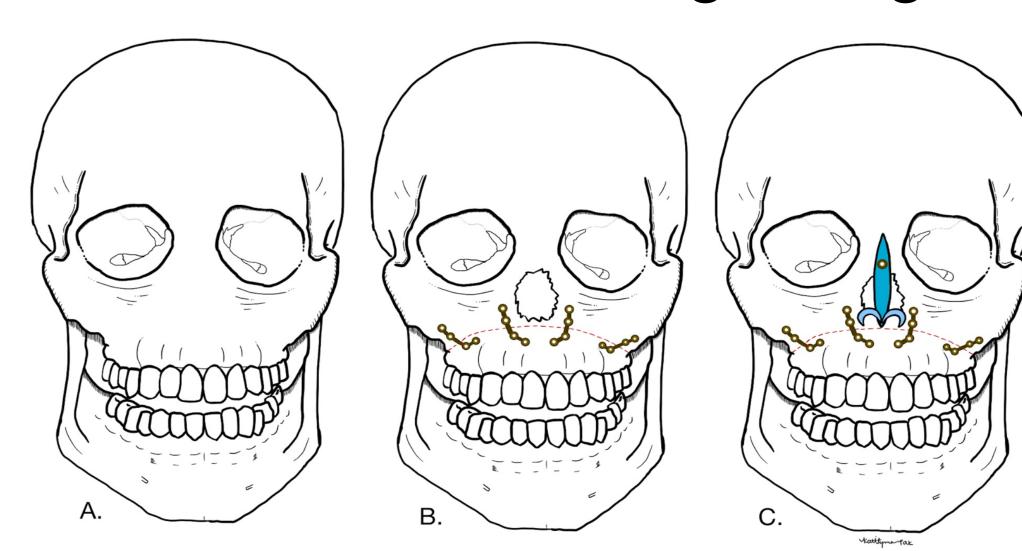


Multi-Staged Reconstruction

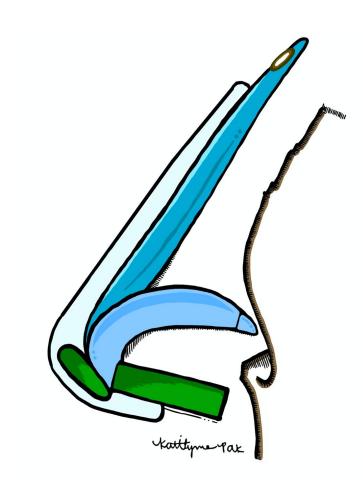
- 1. CT-based stereolithography + prosthesis placement (age 11
- 2. Methylmethylacrytate prosthesis adjustment (2 stages, ages 13 & 15)

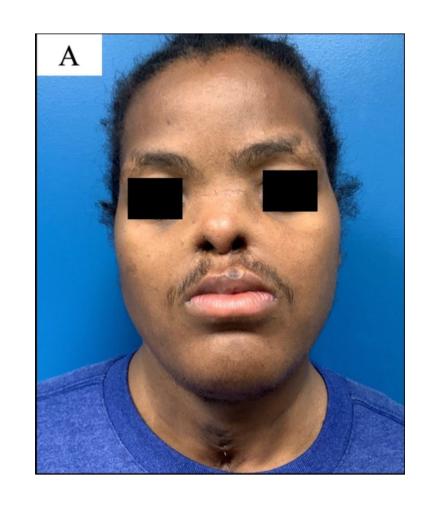


- Decannulation and airway reconstruction (age 22)
 - -Internal maxillary artery embolization
 - -Lefort 1 osteotomy (9 mm advancement)
 - -Burring posterior segment
 - -Full thickness skin grafting of nasal airway



- 4. External nasal reconstruction (age 22)
 -Shield tip graft, columellar, dorsal struts
 - -Bilateral conchal bowl alar grafts





Case Correlates

- 43 cases reviewed
- 4 cases linked to chromosomal/ genetic aberrations, potential AD/AR inheritance
- Associated physical features:

Total # Patients
20
13
10
17
13
2
12
4
6

Conclusions

- Multi-staged reconstruction w/ stereolithography, methylmethylacrytate prosthesis, and combined shield tip graft, columellar & dorsal struts, and bilateral auricular alar grafts is a viable approach to complete arrhinia
- 2. Reconstruction can be accomplished at a later age vs. traditional preschool years
- 3. Pros: limited scar burden, eliminates need for fore-head based or free flaps
- 4. Cons: multiple procedures, potential loss to follow-up

References

Ali et. Al 2014; Fernandes et. Al 2016; Fuller et. Al 2020; Li et. Al 2015