

Congenital Complete Arhinia: Report of a Rare Case

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Congenital absence of the nose (arhinia) is extremely rare. Arhinia leads to severe airway impairment and poor feeding in the neonate. The degree of nasal absence varies from case to case. Here, we present a case of congenital complete arhinia associated with other craniofacial anomalies. Arhinia leads to severe respiratory distress and feeding problems in newborn warranting emergency management, but our case differs as the child was adapted to mouth breathing and naso-gastric tube feeding, giving us the opportunity to delay the definitive management for a few weeks yet avoiding the risk of operating in a small child to create nasal airway or tracheostomy on an emergency basis.

Keywords: Arhinia, Congenital anomalies, Newborn

INTRODUCTION

Congenital arhinia is an anomaly consisting of the absence of external nasal structures and nasal passages.¹ It is a rare case of anomaly of embryogenesis with only 28 cases have been reported since 1931 in literature.² Here, we present a case of congenital complete arhinia associated with other craniofacial anomalies.

CASE REPORT

A female, term baby was the first child of unrelated parents. The mother had uneventful antenatal history, and the ultrasound examination was within normal limits. On physical examination, it was found that the baby had no external nostrils. Nasal pyramid was absent and there was a palpable bony prominence probably of rudimentary nasal bones covered with normal dermis at the nasal radix area (Figure 1). She had hypertelorism, and the right eye was hypoplastic. There was right pre-auricular tag (Figure 2). Rest of the physical examination was unremarkable. She had no cyanosis, and she was breathing per mouth. On 2nd day of life, she was started feeding via orogastric tube. Computerized tomographic (CT) scan showed rudimentary nasal bones. Nasal passage was absent along-with maldeveloped nasal cartilage, nasal cavity and paranasal sinuses were also noted. Right eyeball, orbit, and right mastoid antrum were also hypoplastic (Figure 3). CT scan of the brain also revealed wide apart lateral ventricles, focal hypodensity in the occipital lobe with cortical sulci and gyri not well-delineated. (Figure 4). Abdominal ultrasound examination and echocardiography were normal. The baby adapted to breathe through mouth during the period she was

admitted in neonatal intensive care unit and hence no surgical intervention for breathing was planned at this stage.

DISCUSSION

The pathogenesis of arhinia is poorly understood. It has been postulated that lack of development of the nose results from failure of the medial and lateral nasal processes to grow, but it is also possible that overgrowth and premature fusion of the nasal medial processes result in formation of an atretic plate. Arhinia may also result from lack of resorption of the nasal epithelial plugs during the 13th-15th weeks of gestation. Another explanation may be related to abnormal migration of neural crest cells to this region, resulting in aberrant flow of the multiple mesodermal structures required to establish the nose and its cavities normally.³ Congenital arhinia is a very rare defect during embryogenesis. Most cases are sporadic, but familial cases have been reported.¹

Olsen and associates reviewed the literature through 2001 and collected 22 cases.⁴ McGlone collected 27 cases until 2003 and investigated the common abnormalities.² Akkuzu *et al.* reported a case of congenital partial arhinia in 2007,¹ and another was reported by Tanyeri *et al.* in the same year.⁵ Thornburg *et al.* reported prenatal diagnosis of total arhinia with normal chromosomal analysis,⁶ and another was reported by Fakhraee *et al.*⁷ Thus, our case becomes the 32nd case of arhinia reported. Most of the collected cases had an uneventful antenatal history and this was true for our case.

Arhinia may be accompanied by variety of anomalies such as absence of the paranasal sinuses, cleft palate, facial anomalies

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Figure 1: Clinical photograph of the baby showing absent external nostril, nasal pyramid, bony prominence at radix with hypoplastic right eye and hypertelorism



Figure 2: Clinical photograph of the baby showing right pre-auricular skin tag

such as hypotelorism or hypertelorism, central nervous system anomalies, umbilical hernia, syndactylia, and hypospadias.^{2,4} Some of these have been found in the case under consideration such as anomalies of paranasal sinuses, right eyeball and orbit, central nervous system anomalies, hypertelorism and hypoplastic right mastoid antrum and right preauricular tag.

The clinical consequences of congenital arhinia are severe airway obstruction and inability to feed. However, some cases have been reported without severe airway obstruction as the child learns mouth breathing from a very early age and so is our case. Most authors agree that surgical reconstruction of the external nose and inner cavities should be delayed at least until preschool years, when facial development is nearly complete.^{8,9}

CONCLUSION

The degree of nasal absence in arhinia varies from case to case. Although we know newborns to be obligate nose breathers, some cases of arhinia may develop an alternative

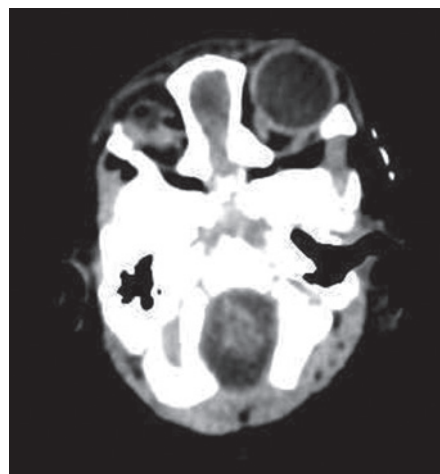


Figure 3: Computerized tomographic scan picture showing rudimentary nasal bones, absent nasal passage with mal-developed nasal cartilage, nasal cavity and para-nasal sinuses. Right eyeball, orbit and right mastoid antrum were hypoplastic

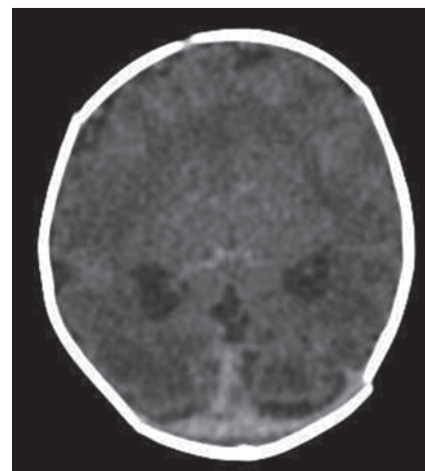


Figure 4: Computerized tomographic scan brain showing wide apart lateral ventricles, focal hypodensity in occipital lobe with not well delineated cortical sulci and gyri

way of breathing through mouth. Thus, they may bypass placement of an oral airway or surgically created nasal airway or a tracheostomy tube in acute cases and later definitive treatment can be planned accordingly.

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