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# The arrhinias: Proboscis lateralis literature review and surgical update

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ABSTRACT

Proboscis lateralis (PL) is a rare malformation, reported for the first time in 1861 by Forster in his monograph on congenital malformations of the human body. The abnormal side of the nose is represented by a tube-like rudimentary nasal structure, attached at any point along the embryonic fusion line between the anterior maxilla and the frontonasal processes. As clefts of the lip (and alveolus) are bilateral or unilateral, an arrhinia can be bilateral (total) or unilateral. In this case it is a 'hemi-arrhinia' (or heminasal agenesis. The arrhinias represent three groups of anomalies, each with different levels of clinical severity, some involving association with the labio-palatal cleft or agenesia of the premaxilla (1). In PL the nasal cavity on the affected side is replaced by a tubular appendage located off-center from the midline of the face, arising commonly from the medial aspect of the roof of the orbit (2). It is usually associated with heminasal aplasia or hypoplasia, microphthalmia, and — less commonly — with midline clefting. Associated brain and cranial vault anomalies are seen in 19% of these patients. PL is usually unilateral, with very few symmetrical/bilateral cases being reported (3). Morpho-aesthetic and psychological problems are frequent concerns for the patients and their families.

In this study, the authors describe a clinical case and the chosen surgical technique, as well as reviewing the alternative techniques present in the literature.



## 1. Introduction

A review of the literature classified proboscis lateralis (PL) into four groups (Khoo BC 1985).

1. Proboscis with normal nose
2. Proboscis with nasal defect only
3. Proboscis with nasal defect, plus defect of eye and adnexa
4. Proboscis with nasal defect plus abnormality of eye and its adnexa plus cleft lip or palate or both.

PL often attaches in the upper medial canthal region (Losee et al., 2004); however, atypically, it can lie in the lateral canthus, the lateral supraorbital ridge, the mid-upper eyelid, the root of the nose in the midline, the chin (van der Meulen et al., 1983), and just above the nostril.

Embryological development of the face and nose is a complex process. The nasal placodes appear to function as the primary organizers for the developing nose. They invaginate into the frontonasal process, separating this structure into the medial and lateral nasal processes. The precise embryological mechanism responsible for the development of PL has not been defined. Popular theories include imperfect fusion of the lateral nasal and maxillary processes, and aberrant fusion of the maxillary process of the affected side to the medial nasal process (globular process) (Harada and Muraoka., 2001; Tessier et al., 2009; Rosen and Gitlin, 1959).

For the complete evaluation of this anomaly, a CT scan is important because it allows assessment of growth of facial and skull bones, as well as CNS development.

Management should start early in childhood to avoid the psychosocial consequences related to this deformity. Complete morpho-aesthetic outcome is delayed until late teens, when growth of the nasal skeleton is almost complete. Cartilaginous or bony support can be planned later in teenhood.

Although many surgical techniques have been described for reconstructing PL, there are two eminent approaches. According to

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Denecke and Meyer (1967), it was Young in 1949 who was the first to mention the correction of the proboscis in the literature. In this approach, the medial or distal half of the tube is de-hepitelialized and sutured to the split lateral wall of the ipsilateral nose. The other frequently used technique is the 'tunneling' method. According to Denecke and Meyer, this method was first reported at the French convention for plastic surgery in 1956 by Recamier and Florentin. In this case, the tube is totally de-hepitelialized except for the distal segment, which is used to form the new ala. It is then brought to the area of the missing nostril through a subcutaneous tunnel formed on the lateral wall of the defect side of the nose.

## 2. Clinical case

A 7-month-old male patient from Romania was referred to our unit.

The parents reported an uneventful pregnancy and delivery at the 36th week with caesarean section; a single umbillical artery at birth was reported by the mother. After birth, growth and breast-feeding were regular.

The patient was initially hospitalized in the pediatric intensive care unit of our hospital, where a general assessment was carried out.

Blood tests suggested a renal insufficiency; a subsequent renal ecography showed the absence of a right kidney. Scintigraphy confirmed the right kidney agenesis and a left compensatory hyperactivity.

The patient presented with hemiarrhinia, with a normal left nostril and columella and total absence of the right side of the nose. On the right side was a tubular structure with cranial base at the level of the glabella, with no communication with airways Fig. 2 A-C. With palpation it was possible to note the presence of a rudimentary right nasal bone at the base of the proboscis. Other features included palpebral fissure length asymmetry, epicanthal folds, normal eyebrows and eyelashes, and low set ear. The lacrimal system was apparently normal, with no signs of obliteration or epifora. No cleft lip or palate, or other significant dysmorphisms, were noted.

A craniofacial CT scan and MRI confirmed the clinical findings, with a bony wall at the right nasal fossa without any communication with the rest of the nose.

On the basis of the peculiar malformation observed, the association with right renal agenesis, and the single umbilical cord reported at birth, a genetic evaluation was performed with array CGH analysis to verify whether genetic anomalies were present to explain the reason for the deformity. The array used was CytoChip Oligo ISCA 8 × 60 K, Bluegnome Ltd.

After discussion and consultation among the authors, a staged surgical repair was planned. The first operation consisted of a translocation of the proboscis towards the left nostril in order to create as normal a nose as possible.

The operation was performed under general anesthesia, starting with the incision of the medial part of the proboscis and raising of a flap comprising 2/3 of the proboscis. The pedicle of the proboscis was based in the superolateral portion.

Incision of the recipient site underneath the proboscis was followed by subperiosteal dissection of the osteocartilagineous portion of the nasal bones, which appeared deformed and medially

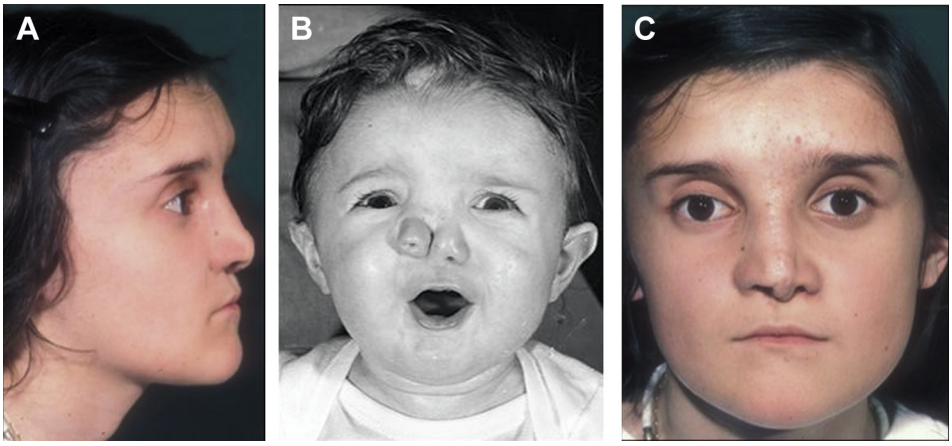


**Fig. 1. A,B,C**: Original Tessier's reconstructive protocol: case of PL and pseudo nasal duplication shown before and after multi step nasal reconstructions also with forehead flap. Paul Tessier 's case. (courtesy Dr. S. A. Wolfe, Miami USA).

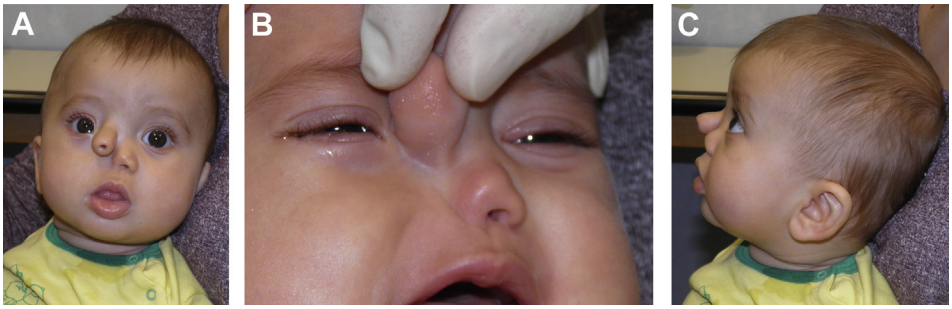


**Fig. 2. A,B,C**Right PL 3-cm long and 1-cm diameter soft trunk like process associated with right nasal fossa disembriogenesis, no air passage, a slight degree of telecanthus.

and upwardly displaced. With the use of a piezosurgical device, the displaced right nasal bone was removed and subcutaneous tissue defatting was carried out.

On examination, the right nasal fossa was found to have a dead end, with no airways communicating with the pharynx.

The flap was repositioned parallel to the left nostril and sutured on top of the recipient site, opposing the two de-hepitelialized sites Fig. 3 A-C. The patient was discharged from the hospital 5 days postoperatively.

The second operation was performed 8 months later. Under general anesthesia, a 12 × 8 mm auricular cartilage graft was harvested and used for reconstruction of the nasal tip. The operation started with an incision of the columella, followed by dissection and separation of the skin from the underlying cartilages at the tip of the nose and nasal dorsum. The cartilage was trimmed and grafted on the tip of the nose, and sutured with transfixed stitches to the dorsum, creating an increased nasal tip projection. Small fragments of cartilage inserted in the columella as columellar struts.

An incision was made at the base of the alae, and a transfixed stitch suture used to achieve alar base width reduction. Before suturing, an incision along the scar on the right lateral portion of the nasal tip was carried out to excise residual scarring skin in order to correct and reduce the previous scar.

Sutures and placement of nostril conformers anchored with 5.0 silk sutures completed the operation. The patient was discharged from the hospital 4 days postoperatively and followed up at 6 months up to present Fig. 3D.

## 3. Results

The result of genetic evaluation was Arr (hg19) (1e22) 2,(xy) 1, with no segmental anaeuploides, eliminating the presence of interstitial microdeletion and microduplication (mean resolution of 150_250 kb) as a cause of the peculiar phenotype presented. This meant that there was no known genetic anomaly as the basis of the deformity.

After the first operation the result was a more natural nasal shape, with a single tip and a slight furrow on the midline.

After the second operation and one year follow-up, the result was a good nasal shape and projection with some residual asimmetries. (Fig. 4 A,B,C). In the future other operations are planned in different stages.

## 4. Discussion

Proboscis lateralis is a rare congenital anomaly, with an incidence of about 1 in 100 000 live births, without gender preference, although a study by Boo-Chai reported a 2:1 male-to-female ratio.

Although PL has been described in the absence of other congenital anomalies, it is characteristically accompanied by ipsilateral heminasal hypoplasia or aplasia, and rarely by choanal atresia (Chauhan and Guruprasad, 2010). Nasal air flow is almost normal through the single nostril. The maxilla develops poorly due to the lack of pneumatization of the maxillary sinus, the unerrupted canine (or canines), and the septo-vomerian septation, which is misplaced to the affected side. The obliteration of the naso-lacrimal conduct is inconstant (Tessier et al., 2009).

Ocular and/or ocular adnexal anomalies, as well as cleft lip and/or palate, are most common seen in conjunction with PL. (Chauhan and Guruprasad, 2010).

According to DeMyer, PL can also be seen in association with holoprosencephaly (DeMyer et al., 1964).

As proposed by Tessier, PL can be included under the arrhinias, which are classified into three groups: total arrhinia, hemiarrhinias and proboscis lateralis.

Proboscis lateralis has been divided into four groups by Boochai: Group1, proboscis with normal nose; Group 2, proboscis with nasal defect only on the same side; Group 3, proboscis with ipsilateral nasal defect associated with eye or adnexal defects;

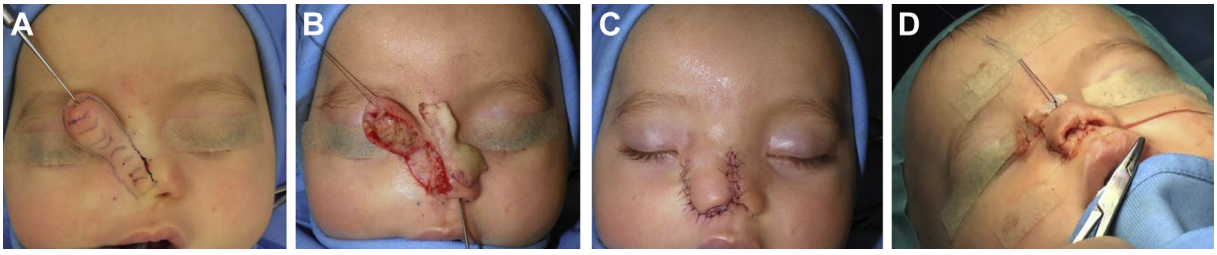


**Fig. 3. A.B.C:** First operation at age 8 months. Splitting and moving of the PL.**D** Second operation at age 15 months.

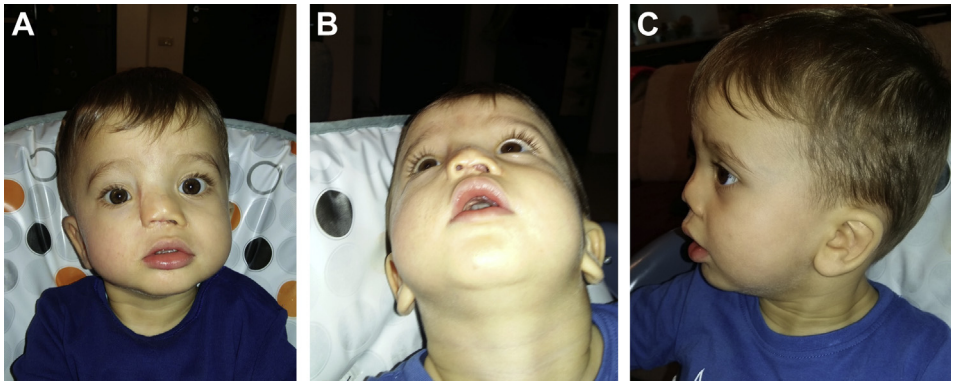


**Fig. 4. A,B, C**: The patient after the two surgeries.

Group 4, in addition to the nasal and ocular defect, a cleft lip or palate.

Prenatal diagnosis of PL has been documented in few cases (Guerrero et al., 2001). The diagnosis is based on clinical examination; radiographs and computed tomography are complementary in determining the extent of the bony and soft tissue components of the anomaly (Chauhan and Guruprasad, 2010).

Although most patients with PL do not have serious central nervous system abnormalities, PL can coexist with central nervous system anomalies, and early neuro-imaging is indicated to rule out intracranial abnormalities (Meeker and Aebli, 1947).

There are few reported cases of PL in the literature, and there is no consensus regarding the timing of surgery and protocols for treatment (Franklin 1921). Tessier suggested that the optimum age should be adolescence, in particular for total arrhinias, starting no earlier than 6–8 years of age. (Tessier) Fig. 1 A-C.

Because there is some variability in facial anomalies and the degree of nasal hypoplasia seen with PL, management must be individualized (Kolluru and Coumary, 2015).

Surgery for PL could be performed earlier, with a different surgical strategy. Some authors suggest removing the proboscis, performing reconstruction with local tubed flaps, and then bone grafting at a later stage (Khoo, 1985). Recaimer and Florentin in 1956 described the tunnelling method. The de-hepitelialized proboscis is passed under a transverse pedicled flap prepared from the lateral skin of the defective nose. In this first stage the nostril is formed. The deformity caused by the pedicle is revised in a second stage.

To reconstruct the deformed heminose, the proboscis is an ideal donor, in preference to the forehead or the upper arm. It is a natural tube pedicle with a good color and texture match, close proximity, and a liberal blood supply. It can be decorticated, leaving a cuff of skin at the margin of the dimple, and then buried subcutaneously alongside the normal heminose in a tunnel or under a flap. Alternatively, the proboscis can be selectively decorticated and attached along its ventromedial aspect to a similar raw area cut alongside the normal half-nose. The groove on the dorsum is smoothed over with a Z-plasty. The third and most popular method is to split the proboscis along its entire length, in a variety of patterns, and attach the resulting convex disk to the normal side of the ala (Khoo, 1985).

## 5. Conclusion

The treatment of proboscis lateralis is still a debated issue. The classical Tessier protocol of treatment starts at an earlier age and follows the baby during growth, even if a long-term follow-up is required. For the case reported here, the authors proposed a modification of the classical Tessier protocol, but also starting at an early stage and following the patient during growth.

Whatever the choice of treatment, the goal of surgery is to create a normal nose, in order to have an acceptable appearance and a better chance of social integration, and to create a functional airway tract when the anatomical structures make this possible. Correct timing and sequencing of reconstruction stages are essential.

In the authors' opinion, reconstruction should be started as early as possible, whilst also considering the psychological aspects for the patient and family.

## Conflicts of interest

None.

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