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Review Article

International Pediatric Otolaryngology Group (IPOG) consensus recommendations: Diagnosis, pre-operative, operative and post-operative pediatric choanal atresia care

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ABSTRACT

Objective: To provide recommendations to otolaryngologists and allied physicians for the comprehensive management of young infants who present with signs or symptoms of choanal atresia.
Methods: A two-iterative delphi method questionnaire was used to establish expert recommendations by the members of the International Otolaryngology Group (IPOG), on the diagnostic, intra-operative, post-operative and revision surgery considerations.
Results: Twenty-eight members completed the survey, in 22 tertiary-care center departments representing 8 countries. The main consensual recommendations were: nasal endoscopy or fiberscopy and CT imaging are recommended for diagnosis; unilateral choanal atresia repair should be delayed after at least age 6 months whenever possible; transnasal endoscopic repair is the preferred technique; long term follow-up is recommended (minimum one year) using nasal nasofiberscopy or rigid endoscopy, without systematic imaging.
Conclusion: Choanal atresia care consensus recommendations are aimed at improving patient-centered care in neonates, infants and children with choanal atresia.

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1. Consensus objectives

To provide recommendations for the comprehensive management of young infants who present with signs or symptoms of choanal atresia.

Choanal atresia is characterized by blockage of the posterior part of the nose, usually by abnormal bony or soft tissue due to failed recanalization of the nasal fossae during fetal development [1]. According to the literature, bilateral choanal atresia accounts for 30% of patients, while unilateral choanal atresia accounts for about 70% of cases [2]. Bilateral choanal atresia is a condition that often requires early surgery because newborns are obligate nasal breathers, while unilateral atresia may be asymptomatic and discovered coincidentally during neonatal examination by the inability to traverse the nasal fossa with a catheter, or even later with unilateral nasal obstruction and discharge [3].

2. Target population

All pediatric patients with signs concerning for choanal atresia, with special focus on the first two years of life.

3. Intended users

These consensus recommendations are intended to inform:

1. Otolaryngologists who perform choanal atresia surgery
2. Allied physicians, including intensive care unit specialists and pediatricians, who collaborate in the care of these patients.

4. Methods

Consensus guidelines based on the review of the literature and expert opinion by the members of the International Pediatric Otolaryngology Group (IPOG). A two-iterative delphi method questionnaire was used to establish expert recommendations on the diagnostic considerations, intra-operative considerations, post-operative considerations and revision surgery. An online survey was designed by two authors of the group (EM and RN). The survey was distributed to all members of the IPOG, and responses were collected. The responders were given the opportunity to comment on the content and the format of the survey, which was modified for the second round. To reflect the variability in practice patterns present among experts in the field, the degree of consensus was quantified by presenting the percentage of above authors who agree with each statement.

5. Disclaimer

Members of the International Pediatric ORL Group (IPOG) prepared this report. Consensus recommendations are based on the collective opinion of the members of this group. Any person seeking to consult this report or apply its conclusions to patient care is expected to use independent medical judgment in the context of individual patient and institutional circumstances.

6. Recommendations and justification

Twenty-eight members completed the survey, in 22 tertiary-care center departments representing 8 countries including Australia, Canada, France, Ireland, Italy, Portugal, the United Kingdom, and the United States of America. The results of the survey are summarized in Table 1 and outlined in the following subheadings:

6.1 Pre-operative considerations
6.2 Intra-operative considerations
6.3 Post-operative considerations
6.4 Revision Surgery

Table 1
Results of the survey about choanal atresia.

	All patients or Yes (%)	In selected cases (%)	Never or No (%)
Initial clinical examination			
Mirror or paper examination under the nostril	39.3	35.7	25
Insertion of a nasogastric tube	10.7	53.6	35.7
Nasofiberscopy/endoscopy	85.7	14.3	0
Preoperative assessment			
CHARGE assessment	35.7	64.3	0
Genetics consultation	35.7	64.3	0
Audiologic assessement	57.1	32.2	10.7
Cardiologic evaluation	25	75	0
Ophtalmology consultation	25	75	0
Urology consultation	3.6	96.4	0
Neurology consultation	7.1	92.9	0
Imaging			
Craniofacial CT-scan	100	0	0
Cerebral MRI	0	100	0
Transthoracic ultrasounds	17.9	82.1	0
Genito-urinary ultrasounds	14.3	85.7	0
Surgical technique			
Endoscopic evaluation	89.3	10.7	0
Transpalatal approach	0	92.9	7.1
Reapplication of mucosal flaps (Yes/No)	35.7	0	64.3
Mitomycin-C	82.1	14.3	3.6
Stenting in unilateral choanal atresia	14.3	60.7	25
Stenting in bilateral choanal atresia	32.1	53.6	14.3
Postoperative management and follow-up			
Proton-Pump Inhibitors (Yes/No)	64.3	0	35.7
Oral or intravenous antibiotics (Yes/No)	46.4	0	53.6
Oral or intravenous steroids (Yes/No)	28.6	0	71.4
Nasal humidification by aerosols (Yes/No)	32.1	0	67.9
Nasal topical steroids (Yes/No)	71.4	0	28.6
Nasofiberscopy for follow-up (Yes/No)	96.4	0	3.6
Postoperative control under GA	25	50	25

6.1. Pre-operative considerations

The members of the IPOG identified important preoperative considerations. Variations in practice among the current group members remain and the purpose of this section is to provide a list of reasonable options based on expert opinion. A summary of the answers is provided in Table 1.

6.1.1. Diagnosis

Each patient with clinical symptoms should undergo a nasofiberscopy or a nasal rigid endoscopy in clinics to confirm the diagnosis of choanal atresia (85.7% of the members of the IPOG). Documenting airflow using a mirror or a paper under the nostrils, at least in selected cases, remains a common test (75%), as well as the insertion of a nasogastric tube in the nasal cavity (64.3%). The choice of the clinical diagnostic tool is guided by its availability, nasofiberscopy or nasal rigid endoscopy elicited strong consensus as they allow direct visualization of the atretic plate and nasal cavity and may aid in surgical planning.

Each patient should undergo CT imaging in order to confirm the diagnosis and plan surgery (100%) [4]. Ideally, slices of the middle and inner ear should be included to look for abnormalities [5]. Nowadays,

CT-scanning is quickly acquired and often does not require general anesthesia.

6.1.2. Associated anomalies evaluation

Choanal atresia is often present in CHARGE syndrome, in which case it may be associated with coloboma, heart abnormalities, retardation of growth and development, genito-urinary abnormalities and ear abnormalities or deafness [6]. In some cases, choanal atresia is associated with abnormalities that are not in the CHARGE spectrum [7]. A major challenge is to detect these associated abnormalities, which can be life-threatening during general anesthesia. Most of the members of IPOG do not systematically perform a complete CHARGE evaluation but rather selectively refer the patients based on risk factors and clinical examination (64.3% vs 35.7%). Nevertheless, most of the members of the IPOG wait for the results of cardiac ultrasound before the surgery for bilateral choanal atresia (78.6%). Most of the members of IPOG selectively request a genetics consultation (64.3%), not systematically (35.7%).

An audiologic assessment should be performed in all patients with choanal atresia (57.1%), using the adapted method in function of the age at diagnosis and of the clinical presentation.

6.1.3. Timing for surgery

For unilateral choanal atresia, surgery should be delayed after the age of at least 6 months (92.8%), ideally one year (78.5%) or two years (36%) (Fig. 1). The emergence of symptoms may lead to earlier surgery (Fig. 2).

For bilateral choanal atresia, which leads neonatal respiratory distress, most of the members (78.6%) wait for a short assessment of the risk factors for general anesthesia, mainly cardiac abnormalities (Fig. 3), but others perform it just after diagnosis (24.1%).

6.2. Intra-operative considerations

Transnasal endoscopic repair for choanal atresia is the preferred initial technique with transpalatal approach reserved for patients in whom transnasal repair would be impossible (82.1%). Transnasal repair is usually considered as impossible when the nasal fossa does not allow the simultaneous use of endoscope and instrument [8]. No member of the IPOG reported systematically using the transpalatal approach as initial treatment and some members never (7.1%) or almost never (7.1%) utilize this technique. There is no cut-off weight to choose whether to do a transpalatal or a transnasal repair; the choice depends on the local anatomy (89.3%). Use of nasal endoscopy is recommended

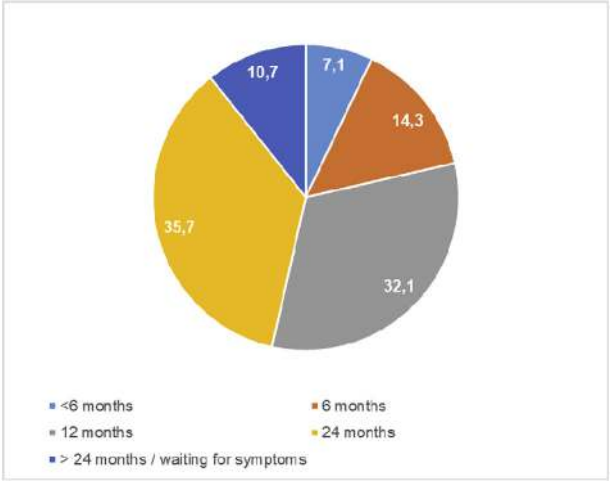

Fig 1. Ideal timing for unilateral choanal atresia repair. Most of the members of the IPOG delay surgery after 6 months (92.6%), and ideally after One year (78.5%).

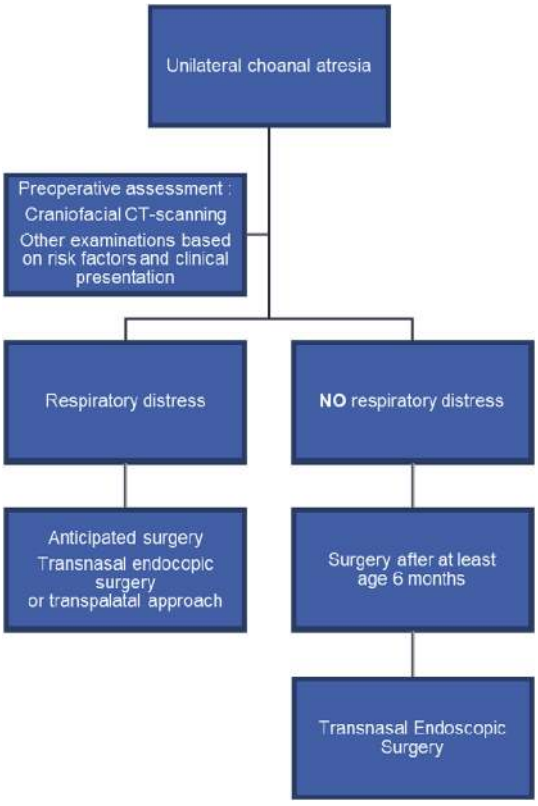

Fig. 2. Management algorithm for unilateral choanal atresia.

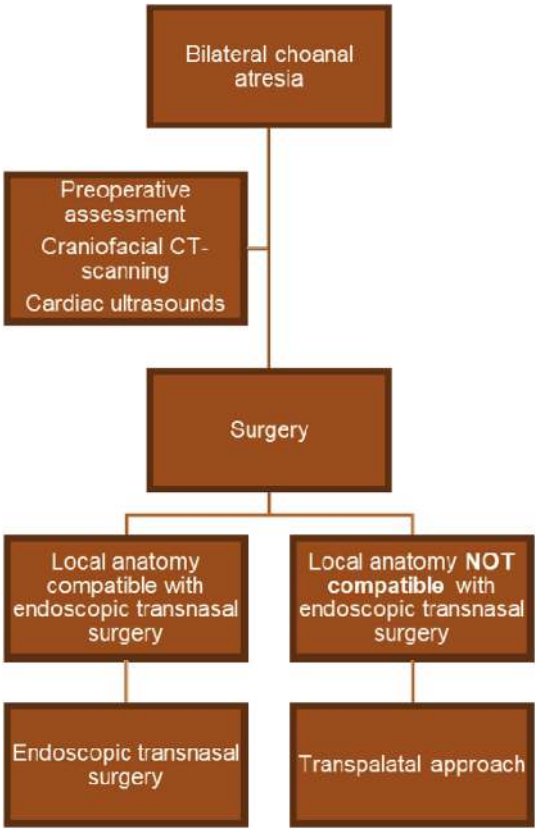

Fig. 3. Management algorithm for bilateral choanal atresia surgical repair.

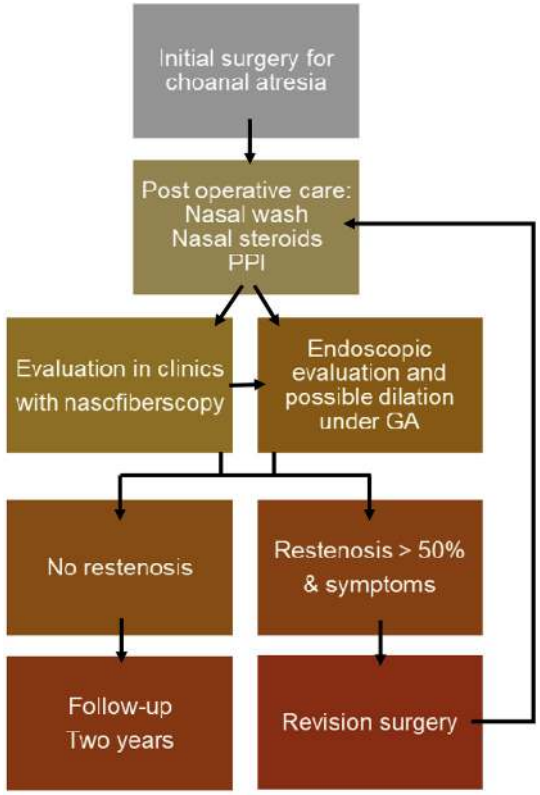

Fig. 4. Post operative decision algorithm after choanal atresia surgery.

(89.3%) during the surgical procedure for choanal atresia, even in the event of transpalatal repair.

The preferred endoscopic technique is highly surgeon-dependent. Drilling of the atretic plate and mucosal microdebridement are the most commonly performed, but balloon dilation and cold instruments are often performed. Laser has been cited by one member. Reapplication of mucosal flaps is not commonly performed: 35.7% of the members re-apply flaps, 64.3% do not.

Mitomycin-C is not to be used in first intention (never for 82.1%). Stenting is less frequently performed than in the past and is considered necessary by most respondents (60.7%) only in selected cases. The preferred stenting material is an endotracheal tube (75%), but nasal trumpets, chest tube or Silastic sheets have been cited.

6.3. Postoperative considerations (Fig. 4)

Proton-pump inhibitors are recommended by 63.4% of the members of the IPOG, with usual duration of prescription less than two months (57.1%). The prescription of antibiotics is debated: 46.4% of members prescribe oral or intravenous antibiotics, 53.6% do not. Intranasal corticosteroids are commonly used (71.4%) while systemic corticosteroids are not (28.6%).

Nasal saline should be applied after surgery (100%), with a duration less than or equal to four weeks recommended by 82.1% of members. Frequency of nasal wash varying between one to three times a day is recommended (75% of members).

Clinical follow-up should include nasofiberscopy or nasal rigid endoscopy (96.4%). The timing of the first postoperative control is in the first two weeks for 75% of members, between two and four weeks for 25%. A second look under general anesthesia is performed in selected patients by 50% of members and systematically by 25%. Reasons for preemptive post-operative evaluation under GA are bilateral choanal atresia, syndromic choanal atresia (like CHARGE), low weight, low age, and the surgical technique. The majority of members (64.2%) report the timing of this intervention to be within the first month after initial surgery.

Postoperative imaging is not recommended, even in case of revision surgery (96.4%). Objective assessments of nasal breathing are rarely performed and rarely feasible in this population of newborns and infants.

The duration of the follow-up should be long, at least one year (100%): one year (20%), to adulthood (33.3%), two years being the most consensual (46.7%). The surgical result is considered as stable after 6–12 months (73.3%), but delayed restenosis has been described and this information has to be given to the parents and the patients.

6.4. Revision surgery

Revision surgery should be performed when breathing difficulties linked with re-stenosis are present (82.1%). Another argument to indicate revision surgery is nasal discharge with stenosis.

Endoscopic techniques were preferred in revision surgery, with the same associations of instruments (drilling, microdebrider, balloon dilation). There is no consensus for the use of Mitomycin-C. 42.9% of members recommended its use in refractory choanal atresia, but 32.1% report never using it. Stenting is more commonly used in revision surgery than in initial surgery, using the same materials, and should be performed based on intra operative findings.

7. Conclusion

Choanal atresia care consensus recommendations are aimed at improving patient-centered care in neonates, infants and children with choanal atresia. Diagnosis is made by the combination of nasofiberscopy or nasal rigid endoscopy and CT imaging. Unilateral choanal atresia repair should be delayed after at least age 6 months whenever possible. Transnasal endoscopic repair is the preferred technique for bilateral and unilateral choanal atresia whenever possible. Long-term follow-up is recommended (minimum one year) using nasal fiberscopy or rigid endoscopy, without systematic imaging.

Conflicts of interest

None.

Acknowledgments

Drs Richard Nicollas (senior author) and Eric Moreddu (first author) were the lead authors and Dr Mark Rizzi provided primary consulting and guidance regarding the design and the wording of the consensus recommendations. All remaining authors are listed in alphabetical order. The authorship list follows the agreement of the members of the IPOG. All authors have contributed to the conception and the design of the work, drafting and revising the consensus recommendations for important intellectual content, final approval of the version to be published, and agreement to be accountable for all the aspects of the work.

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