Acta Otorrinolaringológica Española 2021;72(1):51–56

Acta Otorrinolaringológica Española

www.elsevier.es/otorrino

REVIEW ARTICLE

Performance of endoscopic repair with endonasal flaps for congenital choanal atresia. A systematic review

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Received 14 July 2019; accepted 19 January 2020
Available online 18 May 2020

KEYWORDS
Endoscopic CAA repair;
Choanal atresia;
Endonasal flaps atresia;
Endonasal flaps atresia repair;
Choanal atresia repair

Abstract Endoscopic repair of congenital choanal atresia is the gold standard surgical treatment today. Though several controversies on treatment have been reported, surgical techniques for better outcomes are still in discussion. The objective of this study is to evaluate the performance of endoscopic choanal atresia repair with endonasal flaps and no stents.

Publications in English in the last 5 years were searched in the PUBMED database and were systematically reviewed. A total of 9 articles were included according to the inclusion criteria, obtaining a total of 266 patients managed for congenital choanal atresia with endoscopic surgery, endonasal flaps, and no stents. Surgical results, type of atresia, atresia laterality, associated pathologies and follow up were evaluated.

Successful surgery was obtained in 237 (89%) patients while 29 (11%) patients required a new surgical intervention during the follow-up period. Fourteen percent of the patients were diagnosed with CHARGE syndrome and 5% of the patients had some associated heart disease. Bony-Membranous stenosis was observed in 74% of the patients, while a total bony obstruction was recognized in 26% of the patients. Unilateral atresia was observed in 37% of the cases and 63% of the cases had bilateral atresia. The mean follow-up period was 39.5 months (range 3–168 months).

An important functional success rate can be accomplished by correcting congenital choanal atresia using functional endoscopic surgery, covering raw areas with endonasal vascularized flaps, avoiding postoperative endonasal stenting.

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https://doi.org/10.1016/j.otorri.2020.01.002

PALABRAS CLAVE
ACC cirugía endoscópica;
Atresia de coanas;
Colgajos endonasales;
Reparación atresia coanas con colgajos nasales;
Reparación atresia de coanas

Rendimiento de reparación endoscópica de atresia de coanas con colgajos endonasales. Revisión sistemática

Resumen La reparación endoscópica de la atresia de coanas es hoy en día el tratamiento de elección. Existen controversias con respecto a la técnica quirúrgica que aporte los mejores resultados. El objetivo de este estudio es evaluar el rendimiento de la reparación endoscópica de la atresia de coanas con uso de colgajos intranasales, sin uso de *stents*. Se realizó una revisión sistemática de los artículos escritos en inglés publicados en la base de datos de PUBMED en los últimos 5 años. Un total de 9 artículos cumplieron los criterios de inclusión, reuniendo 266 pacientes que fueron tratados de una atresia de coanas con técnica endoscópica con colgajos intranasales, sin uso de *stents*. Las variables evaluadas fueron: el tipo de atresia, los resultados quirúrgicos, la lateralidad, la enfermedad asociada y el seguimiento. En 237 pacientes (89%) se consiguió un resultado satisfactorio, mientras que 29 pacientes (11%) requirieron una nueva intervención quirúrgica durante el seguimiento. El 14% de los pacientes fueron diagnosticados de síndrome de CHARGE y un 5% tuvieron alguna cardiopatía asociada. Una estenosis óseo-membranosa fue encontrada en un 74%, mientras una estenosis totalmente ósea fue observada en un 26%. Lesiones unilaterales fueron observadas en el 37% de los casos y bilaterales en el 63% de los casos. El seguimiento medio fue de 39,5 meses (rango: 3-168 meses). Un éxito quirúrgico funcional se puede obtener usando cirugía endoscópica nasal, cubriendo las zonas de exposición ósea con colgajos nasales vascularizados y evitando el uso de *stents* postoperatorios.

Introduction

Congenital choanal atresia (CCA) is defined as a congenital obstruction of the nasopharynx. The most widely accepted hypothesis for its etiology is an abnormal neural crest cell migration. The incidence arranges between 1 in 5000–9000 newborns. Most cases are unilateral and are more frequent in women.[1] Bony-membranous (mixed) obstruction is detected in approximately 70% of patients. Pure bony obstruction is observed in 30% of patients and occurs secondary to an enlarged vomer or medialized pterygoid plates. Congenital abnormalities such as CHARGE syndrome (coloboma, heart defects, atresia of the choanae, retardation of growth and development, genital and urinary abnormalities, ear abnormalities with or without hearing loss) and craniofacial syndromes such as Treacher Collins are usually correlated with bilateral stenosis.[1] Between 20% and 50% of patients with CCA have other associated congenital anomalies or syndromes. Newborns with bilateral congenital choanal atresia are obligate nasal breathers, which produce respiratory distress. Typical signs consist of cyclical cyanosis, which is relieved by crying.[2]

In 1908, the transseptal approach was described, using the submucous septoplasty-like technique. A portion of the septum was resected in-block with the atretic plate in order to create a neo choana. The problem was that the resection of the immature septum caused anomalies of the face and nose development. In 1937 a trans-palatal approach was developed. Large exposure of the choanal region was achieved, enabling drilling of the medial pterygoid plates. Unfortunately, blood loss was a serious complication that arose from this technique. The endoscopic approach was applied to CCA repair in the 1990s, enabling the integration of CCA surgery with the principles of functional endoscopic sinus surgery (FESS). FESS should currently be considered as the gold standard for CCA, and should fulfill the following requirements:

1. Perform the surgery with a minimally invasive procedure.
2. Provide nasal function in a one-stage operation.
3. Minimize tissue damage of neighboring structures
4. Preserve the development of facial structures.
5. Minimize postoperative pain and discomfort.
6. Enable the early start of spontaneous breathing and feeding.
7. Reduce the duration of hospitalization.[3]

Regardless of the access route and repair technique employed, restenosis is the most frequent cause of surgical failure. With the purpose of preventing restenosis and subsequent reoperation, many authors suggest the meticulous preservation of mucosa and use it as vascularized flaps, which occasionally are combined with postoperative stenting. The practice of stenting, however, is controversial given the documented downsides associated with their use; these include physical irritation of mucosa and disruption of the mucociliary function while in situ; potential as a nidus for bacterial colonization leading to persistent inflammation and fibrosis; pressure deformities; trauma to healing mucosa upon stent removal. Although some publications report stenting to have no adverse effects, others have suggested that the omission of stents may reduce the incidence of restenosis by as much as 3 times.[4]

The objective of this review is to evaluate the performance of the transnasal endoscopic repair of congenital

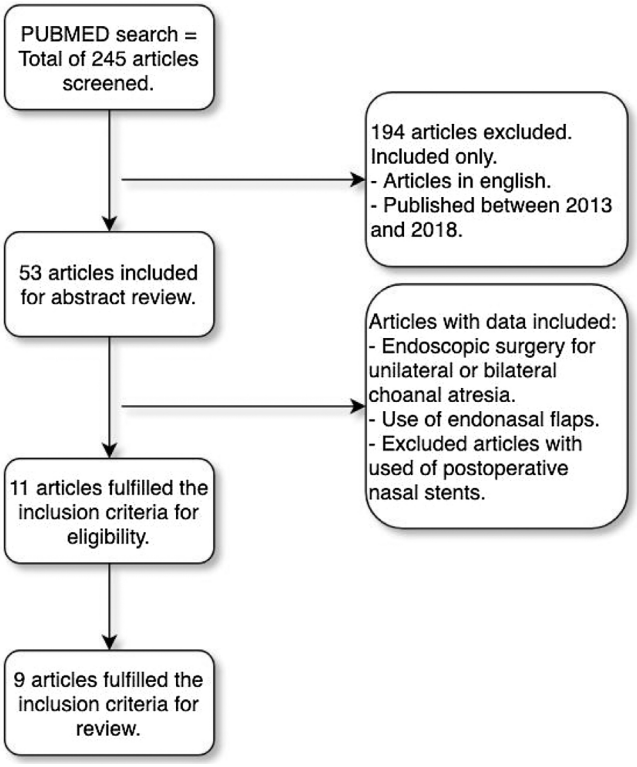

Figure 1 Flow diagram of the systematic literature search and study selection.

choanal atresia using endonasal flaps without the application of stents.

Materials and methods

An extensive search of the literature was performed in the PUBMED database from the last 5 years, from January 2013 to December 2018. Having as primary objective studies with patients who have undergone endoscopic surgery with endonasal flaps for unilateral or bilateral congenital choanal atresia repair.

Using this framework 3 reviewers retrieved studies and were critically appraised. Language restrictions were applied and only articles written in English were included. From a total of 53 articles were evaluated, nine studies continued to meet the defined criteria and were further analyzed. During the search procedure, keywords selected for the study were ''choanal atresia'', ''endoscopic choanal atresia'', ''choanal atresia flap'', ''choanal atresia repair'', ''choanal atresia treatment'', were used. Keywords were either combined with each of the other keywords individually or in groups. Also, references of the retrieved articles were searched. All patients with a history of congenital choanal atresia managed with endoscopic surgery with endonasal flaps were included. Patients that postoperative nasal stents were applied were excluded from the study (Fig. 1).

Tables with analytic data were made using laterality (unilateral or bilateral), Type of atresia (bony or bony-membranous), surgical failure, follow up and the two most frequent associated pathologies described: CHARGE syndrome and associated cardiopathies.

Surgery was considered successful if the patient showed no symptoms of airway obstruction. Objectively, the operation was considered successful if the new choana remained patent post-operatively and the patient was asymptomatic during the follow-up period. If surgery was unsuccessful, a new surgical intervention was required.

Surgical technique: All studies acknowledged that the most critical step for surgical success was the removal of the posterior third of the vomer bone, removal of the atretic plate using as superior margin the projection of the inferior limit of the middle turbinates, as lateral margin the medial pterygoid plate and inferiorly, the palatine bone. Three types of endonasal flaps were used: L-shaped septal flap; inferior and superior based septal flap; and nasoseptal-like flap (Fig. 2).

Results were measured differently depending on the total number of patients gathered for each variable, discarding all patients in the studies where non-available data was found.

Results

Nine studies were appraised and five variables were measured for obtaining results: Associated pathology, type of atresia, laterality, surgical outcome and follow up period. Results are compiled in Tables 1 and 2.

Associated pathology: 4 studies held available data for determining the prevalence of the 2 most frequent associated pathologies in CCA; CHARGE syndrome and cardiopathies. A total of 162 patients were collected, 22 (13.6%) patients were diagnosed with CHARGE syndrome and 8 (4.9%) patients have associated an isolated cardiopathy.

Type of atresia: 8 studies had available data for determining the proportion of atresia type, obtaining 182 patients, bony-membranous stenosis was identified in 73.6% of the patients, while a total bony obstruction was observed in 26.4% of the patients.

Laterality: All studies had available data in relation to if the atresia was unilateral or bilateral. A total of 266 cases were assessed observing 98 (36.8%) cases had unilateral atresia and 168 (63.2%) cases had bilateral CCA.

Surgical outcomes: All studies had available data in relation to surgical outcomes. A total of 266 patients were evaluated. Surgery was considered successful if the patient manifested no symptoms of airway obstruction. Objectively, the procedure was considered successful if the new choana prevailed patent and asymptomatic postoperatively during the follow-up period. If surgery was unsuccessful, a new surgical intervention was required. Successful surgery was obtained in 237 (89.1%) patients while 29 (10.9%) patients needed a new surgical intervention during the follow-up period.

Follow up: 6 studies had available data for determining the follow-up period. A total of 136 patients were evaluated. The mean follow-up ponderated period observed was 39.5 months (range 3–168 months).

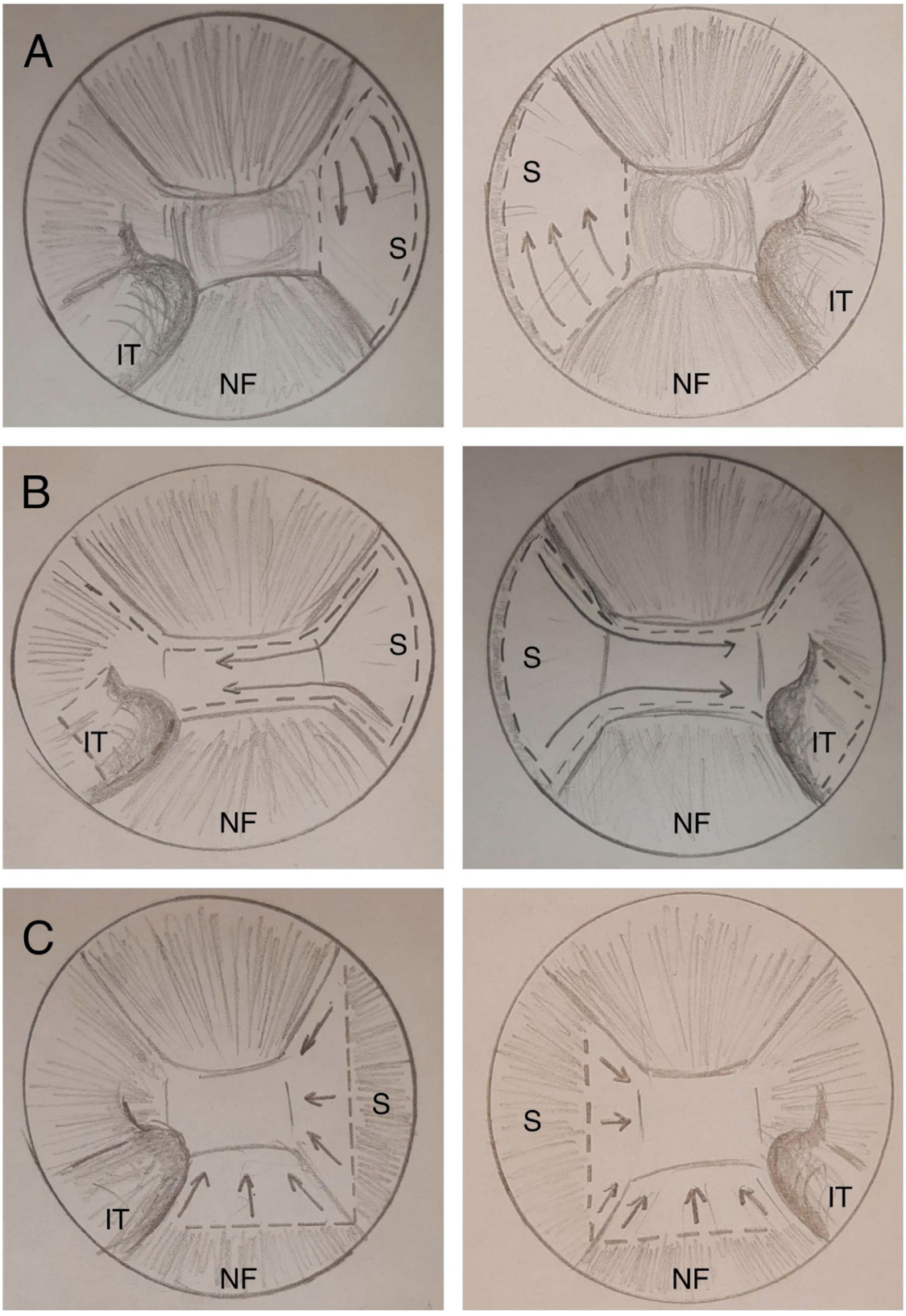

Figure 2 Endonasal flaps description. (A) Image explaining inferior and superior based septal flap. (B) Image explaining nasoseptal-like flap. (C) Image explaining the L-shaped septal flap. IT: inferior turbinate. S: nasal septum. NF: nasal floor. Segmented line and black arrows indicate flap design.

Table 1 Data and results.

Article	*N*	Weighting	Unilateral	Bilateral	Bony-Membranous	Bony	Surgical failure	%	Follow-up (months)	Range
Llorente JL. et al.[5]	10	0.04	3	7	100	0.0	0	0	27.0	11–78
Eladl HM. et al.[6]	29	0.11	0	29	77.7	22.3	6	20.9	NDA	NDA
El-Anwar MW. et al.[7]	25	0.09	0	25	84	16	1	4	35.8	12–66
Wormald PJ. et al.	17	0.06	10	7	82.4	17.6	0	0	NDA	NDA
Brihaye P. et al.	36	0.14	18	18	70	30	2	5.6	72.0	12–168
Gulsen S. et al.[8]	7	0.03	4	3	77.1	22.9	0	0	35.0	3–213
Karligkiotis A. et al.[9]	84	0.32	55	29	NDA	NDA	8	9.5	NDA	NDA
Tatar EC. et al.[10]	16	0.06	8	8	56.3	43.8	3	18.8	37.9	12–105
Tomoum MO. et al.	42	0.16	0	42	63.9	36.1	9	21.4	18.2	8–24
Total	266	1	98	168	73.6	26.4	29	10.9	39.5	3–168

NDA: No data available.

Table 2 Results.

	N	%
Associated pathology		
Total	*162*	
CHARGE	22	13.6%
Cardiopathy	8	4.9%
Type		
Total	*182*	
Bony-Membranous	134	73.6%
Bony	48	26.4%
Laterality		
Total	*266*	
Unilateral	98	36.8%
Bilateral	168	63.2%
Surgical outcomes		
Total	*266*	
Reintervention	29	10.9%
Success	237	89.1%

Follow up	*N*	Months
Total	136	39.5
Range		3–168

Discussion

We have seen in the last years that otolaryngologists have a trend during FESS to use endonasal vascularized flaps for endonasal repairs, and, endoscopic CCA repair is no exception. Over the recent years, new procedures have been developed and the most extensively used is the repair the atretic choanal plate defect endoscopically according to:

1. Removal of the posterior third of the vomer and atretic plate.
2. Superior limit: the projection of the inferior limit of the middle turbinate.
3. Lateral limit: the medial pterygoid plate and inferiorly, the palatine bone.
4. The bony defect is covered with vascularized endonasal flap.

Three methods have been most commonly described: L-shaped septal flap; inferior and superior based septal flap; and nasoseptal-like flap (Fig. 2). One of the major complications of choanal atresia repair is the postoperative restenosis of the neo choana.[1]

The use of mitomycin C as an adjunct warrants discussion. Mitomycin is an antibiotic produced by Streptomyces Caespitosus and has both antineoplastic and antiproliferative properties.[11] It allows for the inhibition of fibroblast proliferation and decreases in scar formation. In a meta-analysis from Strychowsky JE et al. in 2015, two studies reported outcomes associated with the use of mitomycin C. Within these two studies combined, only 7 out of 24 patients (29%) were treated with this topical antibiotic. They concluded that no recommendation about the role of mitomycin C in the treatment of CCA can be made with such limited data. In this same meta-analysis, the primary objective was to determine the outcomes of surgical outcomes with the use of postoperative stents and without them. Concluding that similar success rates for bilateral choanal atresia repair were reported with and without the use of postoperative nasal stents. The use of nasal stents may be associated with more frequent and more severe complications. Perioperative adjuncts such as nasal stenting should be considered in the context of the individual patient and surgeon preference. If the surgeon decides to stent, a duration of 1 week or shorter may be associated with better surgical outcomes.[1]

In another recent review, the authors had concluded that surgical outcomes of CCA repair without stents showed fewer complications, hence, decreasing the intensity of postoperative management, while stent-assisted procedures could be preferred for younger patients with no complications.[12]

Tomoum et al. in 2018 published a prospective randomized trial comparing 2 groups of 72 patients with bilateral CCA, proposing that with the development of less traumatic endoscopic techniques, precise removal of the posterior vomer section, accurate production and preservation of endonasal flaps, and the avoidance of stenting, presents the best chance of success regarding CCA repair.[2]

There are wide variations in success rates for CA repair reported in the literature. The interpretation of success rates and a correlation between various studies is difficult because there is no standard determination of surgical failure. Some authors consider revision surgery the removal of granulation, while others define it as any procedure requiring general anesthesia, including stent replacement or removal.[13] In order to resolve this matter, Karligkiotis A. et al. in 2017 proposed that the absence of symptoms should become the basis of success despite the intranasal anatomical findings. Partial stenosis of any dimension, should not be acknowledged as a failure unless it produces symptoms (functional success with anatomical stenosis). In contrast, an intranasal adequately opened neo choana could be accepted as a failure if it is producing problems (functional failure). The indication for revision surgery is accordingly dictated by symptoms and not anatomical findings.[9] Nowadays success or failure of CCA repair is given concerning its functional results and not merely anatomical findings. It has been proposed that probably the main reason for restenosis after endoscopic repair of CCA is mainly due to granulation tissue from the bone-exposed areas, associated with excessive drilling. Covering these areas with endonasal vascularized flaps should decrease the postoperative granulation and increase functional results.[2]

In a recent review in 2019, the authors had concluded that surgical outcomes of CCA repair without stents showed fewer complications, hence, decreasing the intensity of postoperative management. While stent-assisted procedures could be preferred for younger patients with no complications.[13]

In this review, it was observed that from all 266 patients that were intervened, good functional results were observed in 237 (89.1%), using ESS covering raw bone areas with endonasal vascularized flaps. Results are promising, with time will presumably suit the gold standard for managing this pathology.

Conclusion

The endoscopic approach was applied to CCA and is currently considered as the gold standard management for its repair. A great functional success rate can be accomplished by repairing congenital choanal atresia using endoscopic surgery, covering bone-exposed areas with endonasal vascularized flaps that decrease post-operative granulations, and avoiding the use of postoperative stents.

Compliance with ethical standards

All Authors declare that they have no conflict of interest. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.

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