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Surgical management of Gerhardt syndrome

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Rezumat

Managementul chirurgical al sindromului Gerhardt

Sindromul de imobilizare în adducție a corzilor vocale poate fi produs de paralizia ambilor nervi recurenți – sindromul Gerhardt - sau paralizia întregii musculaturi laringiene intrinseci – sindromul Riegel. Etiologia sindromului Gerhardt este multiplă: chirurgia tiroidei, manoperele de intubație intempestivă, traumatismele cervicale, afecțiunile neurologice, tumorile extralaringiene. Principala manifestare a sindromului Gerhardt este dispneea inspiratorie cu slaba modificare a vocii, datorată paraliziei în poziție paramediană a corzilor vocale cu îngustarea importantă a spațiului glotic. Procedurile chirurgicale de lărgire a spațiului glotic pot fi clasificate în mai multe feluri, principalele lor caracteristici fiind: modificările la nivel glotic, abordul chirurgical clasic cervical sau endoscopic, cu sau fără intervenția la nivelul mucoasei, necesitatea efectuării traheostomiei, echipamentul utilizat. Scopul acestui articol este de a prezenta multiplele tipuri de intervenții utilizate de-a lungul timpului, marcate de dezvoltarea metodelor diagnostice, de anestezie, de echipamentul chirurgical cu instrumente şi tehnologii tot mai sofisticate.

Cuvinte cheie: Gerhardt, dispnee inspiratorie, chirurgie

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Abstract

Adduction bilateral vocal fold immobility syndrome may be due by both recurrent laryngeal nerves paralysis - Gerhardt syndrome - and all intrinsic laryngeal muscles paralysis - Riegel syndrome. Etiology of Gerhardt syndrome is thyroid surgery, intubation's maneuver, trauma, neurological disorders, extralaryngeal malignancies. The manifestations of Gerhardt syndrome are inspiratory dyspnea and slightly influenced voicing by paramedian vocal folds paralysis with an important narrowing of the airway at the glottic level. The surgical procedures for enlargement of the glottic space can be classified in many ways and their major characteristics are: changes at the glottic level; surgical approach: open neck or endoscopic, with or without opening of the mucosal lining; the need for tracheostomy; the equipment used. The aim of this review is to expound the variety of interventions through the last century marked by the development of the diagnostic methods, the anesthesia and the surgical armament with sophisticated instruments and technologies.

Key words: Gerhardt, inspiratory dyspnea, surgery

Introduction

Adduction bilateral vocal fold immobility syndrome may be due by both recurrent laryngeal nerves paralysis - Gerhardt syndrome - and all intrinsic laryngeal muscles paralysis - Riegel syndrome. Gerhardt syndrome is a polyetiologic condition, the leading causes include surgery (predominantly thyroid surgery) – 26 to 59%, intubation's maneuver – 1 to 31%, trauma – 1 to 28%, neurological disorders – 7 to 22%, extralaryngeal malig-

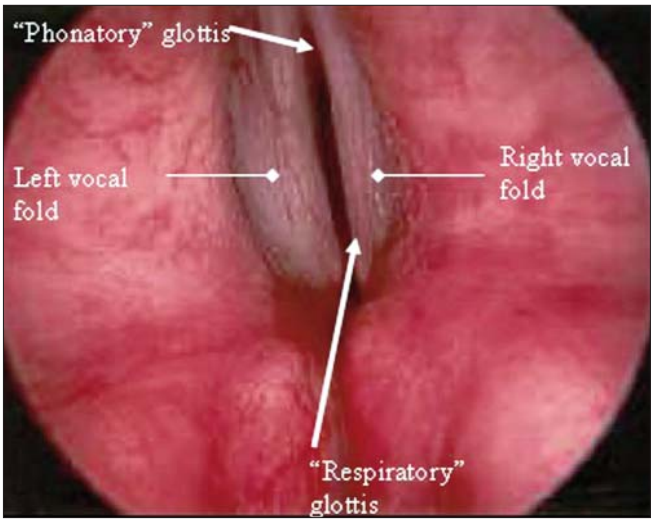

Figure 1. *Endoscopic view of Gerhardt syndrome*

nancies – 5 to 17%. (1) Another clinical entity is bilateral arytenoids cartilage fixation differentiated from bilateral vocal folds paralysis by pre-treatment diagnostic work-up, including electromyography and/or laryngeal endoscopy. (1)

The classical presentation of patients with Gerhardt syndrome is paramedian vocal folds paralysis with an important narrowing of the airway at the glottic level, characterized by inspiratory dyspnea and slightly influenced voicing. *(See Fig. 1)* The condition is life threatening and requires surgical intervention to prevent acute asphyxiation or pulmonary consequences of chronic airway obstruction. Four to fourteen percents of these patients tolerate this condition and do not require any surgical procedure until a viral intercurrent infection decompensate the respiratory status. (2)

For centuries tracheostomy has been the golden standard for securing the airway. A vast majority of surgical interventions have been developed and applied to restore the patency of the airway and achieve decannulation. In the 1950s, the microscopic endoscopic laryngeal surgery boomed. At the end of the twentieth century many of the classical endoscopic operations were performed either with the help of surgical lasers alone, or in combination with other interventions. The surgical procedures for enlargement of the glottic space can be classified in many ways and their major characteristics are: changes at the glottic level; surgical approach: open neck or endoscopic, with or without opening of the mucosal lining; the need for tracheostomy; the equipment used. (3)

The aim of this review is to expound the variety of interventions through the last century marked by the development of the diagnostic methods, the anesthesia and the surgical armament with sophisticated instruments and technologies.

Interventions for Gerhardt syndrome

The major types of interventions include the following: resection of anatomical structures; retailoring and displacing the existing structures, with minimal tissue removal; displacing existing structures, without tissue resection; restoration or substitution of the missing innervations of the laryngeal musculature. (1)

Approaches used for surgery for bilateral vocal fold paralysis are: median thireofissure, lateral transthyroid approach, with a window or after partial resection of the thyroid lamina, paralaryngeal approach, behind the posterior border of the thyroid cartilage, and endoscopic approach.

The arytenoid cartilage is a key structure, the removal of which provides enlargement of the posterior glottis. Some surgeons performed arytenoidectomy in an endolaryngeal way: Iwanoff extirpated the arytenoid through a laryngofissure, with a mucosal incision in the posterior midline of the larynx. (1) Several authors preferred the extralaryngeal way to perform an arytenoidopexy: King done a paralaryngeal approach for mobilized the arytenoid and then it's sutured of the thyroid lamina; Kelly performed the operation through a window in the lower posterior third of the thyroid cartilage; Moritz, Mündnich, Amersbach proposed suturing of the arytenoid to the inferior thyroid corn through a thyroid ala. (2)

The introduction of the endoscopic surgical techniques at the beginning of the second half of the twentieth century was a breakthrough in the laryngeal surgery, including the surgery for bilateral vocal fold paralysis. Kleinsasser played an important role, as he was the first to perform endoscopic laryngeal surgery under microscopic magnification (microlaryngoscopy technique) and to develop special instruments. (3) Thornell performed a small mucosal incision over the arytenoid area extending into the aryepiglottic fold and extract the cartilage. On the basis of Thornell's intervention, Kleinsasser developed his arytenoidectomy and submucosal hemicordectomy. (1)

The introduction of lasers in medicine played a revolutionary role in the development of the modern laryngeal surgery. Its major advantages include: the precise resection, fine cutting line, haemostatic effect, little to moderate postoperative œdema and the absence of cold instruments that could reduce the view field. (4) Disadvantages are thermal trauma, which could induce stenosis and carbonization, which promotes granulations, delayed wound healing and tissue loss and specifically increased anesthesiologic risk with the need for special anesthesiologic equipment and techniques. (4) Ossoff used laser ablation/vaporization arytenoidectomy. An improvement to Ossoff's technique was introduced by Remacle. (4) He proposed the so-called subtotal arytenoidectomy by resection of the body of the arytenoid and preserving only a small posterior shell, which should protect the airway from aspiration. However, the enlargement of the glottis chink may be only moderate unless additional submucosal cordectomy and lateralization are performed.

Several surgical techniques comprise resection of intralaryngeal soft tissues, i.e. parts of the vocal fold, the vocal ligament or the thyroarytenoid muscle. In 1908 Citelli introduced the so-called "chordectomia externa" through thyreofissure (1). Another operation based on resection of the soft tissues and transection of the conus elasticus is the posterior CO_2 laser cordectomy introduced by Dennis and Kashima in 1989. (3) (See *Fig. 2, 3*) Benninger published another technique

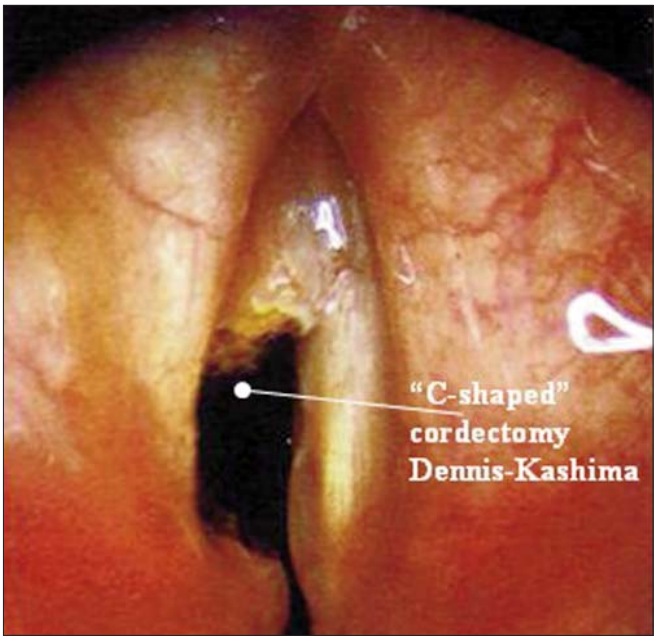

Figure 2. *Intraoperative view: "C-shaped" posterior cordectomy with CO_2 laser*

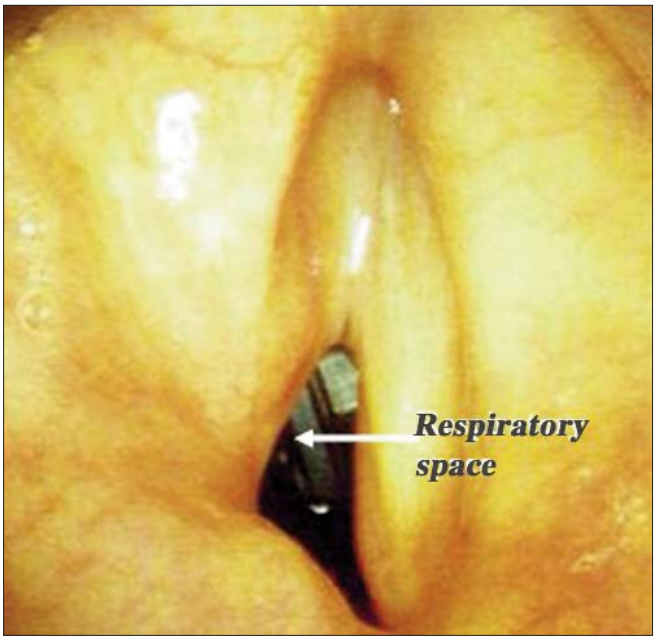

Figure 3. *Postoperative view (after 6 month)*

comprising of laser excision of the posterior part of the vocal cord and submucosal removal of the vocal process with the wound defect being covered with the preserved mucosal flap. (2)

In a large group of surgical interventions the key point is the displacement of the tissues. The majority of them still includes some resection but to a relatively smaller extent if any compared with the first group. The resected structures lie away from the lumen. Réthi performed an operation that is still in use today: the laminotomy with division of the posterior plate of the cricoid cartilage and pulling aside for 5-8 mm with extirpation of the interarytenoid muscle and long-term stenting using a special Réthi stent. (1)

A relatively small group of interventions comprise resection of tissues away from the glottic chink (near the periphery), so the medial structures protruding in the lumen are preserved and only displaced laterally. The objective is to reduce the direct trauma to the phonatory structures and provide better quality of voicing, in addition to preventing scar formation and renarrowing of the lumen. In 1979, Kirchner published his technique of endoscopic longitudinal resection of the thyroarytenoid muscle by means of electrocautery and temporary laterofixation using extra-endolaryngeal suture. (4) Lichtenberger performed a similar intervention with CO_2 laser with endo-extralaryngeal laryngomicrosurgical lateralization include near total arytenoidectomy. (5) In 1991 Kashima reported his early results with transverse cordotomy with CO_2 laser: he performed a transverse incision just anteriorly to the vocal process. (6)

Other techniques are minimally invasive ones where the intralaryngeal structures are displaced without being cut. An important step further was the introduction of the endo-extralaryngeal approach by Lichtenberger, based on a specially constructed needle-carrier.(7) As the atraumatic needles are introduced endoscopically from inside to outside of the larynx, the placement of the threads were done in an extremely precise way.

None of the surgical techniques, based on resection could substitute both major functions of the larynx: enlarging the airway in inspiration and reducing it in expiration for physiologic breathing and phonation. Only restoring the spontaneous and/or voluntary motility of the larynx could achieve such favorable results. The attempts to transpose other muscles to the immobilized arytenoid in order to abduct it and count on spontaneous adduction by the forces of elasticity failed. (8) Crumley performed neuroplastic graft-anastomosis of the recurrent to the phrenic nerve.(9) Tucker performed a reinnervation procedure with the transposition of an active nerve-muscle pedicle (hypoglossal ansa with a small part of the omohyoid or sternohyoid muscle) to the posterior cricoarytenoid muscle. (9)

Discussion

Larynx is a complicated neuromuscular system which simultaneously provides closure for phonation and protection of the lower airways and opening for breathing and cannot substitute by static anatomical remodeling. Only reinnervation procedures constitute physiological approaches to bilateral vocal fold paralysis, but they had proved to be successful in the hands of few surgeons and are currently not applicable in clinical routine.

The tissue resection creates an unphysiologic situation where the phonatory function of the larynx is more or less sacrificed for sake of the airway function. (1) The minimally

invasive methods are preferred to preserve the phonation, perform predominantly in the posterior glottic area and leave the anterior "phonatory" glottis untouched. Endoscopic laser techniques are not superior of any method, but offer the advantage of not requiring a tracheostomy and are therefore extremely well tolerated, with possibility of revision operations. (3)

The most serious complications of the endolaryngeal resection techniques are the scar formation with posterior glottic stenosis and aspiration after arytenoidectomy with lowering of the aryepiglotic fold. The subtotal arytenoidectomy (4) or near total arytenoidectomy with mucosal flap preservation (10) may prevent aspiration.

Bilateral vocal fold paralysis has a relatively low incidence, studies on a larger groups of patients present usually a single technique performed by a senior surgeon. Until a patient gets operated for enlargement of the glottic space, he/she has to be tracheostomised temporarily and live with the stoma for several months. In awaiting period for spontaneous reinnervation electromyography is still uncertain in early diagnosis of reversible and irreversible denervation of the larynx and is unable to select the laryngeal side with better prognosis. Only 2-3 months after could it detect signs of irreversible denervation and thus indicate need for definitive glottal enlargement. (11)

The most important clinical progress at the moment is the avoidance of the temporary tracheostomy by using temporary lateralization procedures. When the indirect laryngoscopy revealed the pathophysiological mechanism of the dyspnea caused by Gerhardt syndrome, physicians became familiar with the fact that in a significant proportion of the patients, spontaneous restitution of the motility could occur after different intervals of time so they could be decannulated. (12) If the recurrent nerves have not been transected during thyroid surgery spontaneous recovery of uni- or bilateral vocal fold mobility could be expected in 40 and up to 86% of the cases. (12)

Recently several new techniques and surgical approaches to the vocal folds have been proposed. These include predominantly anatomic studies, animal experiments or case reports. One of the most promising is Cohen's attempt to paralyze the cricothyroid muscle with botulinum toxin and in this way to reduce its medialization effect on the vocal fold. This intervention was shown to be successful in dogs. (13)

Conclusion

The main treatment modality for adult Gerhardt syndrome is surgery with two major options: tracheotomy and/or glottis enlarging laryngeal surgery. For over a century numerous surgical techniques for glottis enlargement have been introduced, known mainly by their eponyms.

The current trends from the twentieth century are towards endoscopic minimally invasive techniques. Lasers proved to be a very helpful tool for the surgical intervention itself, but it is impossible to state if they are superior to cold steel techniques concerning the late results. Avoiding tracheotomy during the period of waiting for potential spontaneous reinnervation (for example using temporary lateralization procedures) improves significantly the quality of life of the patients and probably will lead to a major change in the overall clinical management of bilateral vocal fold paralysis in adduction.

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