Congenital cricoid stenosis

Pathogenesis, diagnosis and method of treatment

R. N. P. Berkovits, C. E. Bos, K. H. Pauw (Rotterdam)
and A. W. J. de Gee (Appendix) (Apeldoorn, The Netherlands)

Summary

A search of the literature on this subject reveals that in cases of congenital cricoid stenosis an effective method of treatment has yet to be achieved.

In the three cases with this congenital anomaly described here, it was decided to remove the cartilaginous obstruction first. In one case this was done endolaryngeally, in the other two by laryngofissure. Thereafter the patients were intubated nasotracheally, using specially prepared siliconized tubes. In addition, corticosteroids and hyaluronidase were injected into the lesion several times. In all three cases this method of treatment has resulted in an adequate airway. Moreover, a significant impairment of vocal function was not observed.

Historical review

Congenital subglottic stenosis is a rare malformation, discovered mainly by necropsy. Rossi was the first to describe this anomaly in 1826 (Smith and Bain, 1965).

Potter (1962) reported only three cases from over 6,000 neonatal necropsies. All three infants had a solid mass of cartilage, approximately 3 mm thick, occluding the entire larynx immediately below the vocal cords. In two of these cases, a hair-like canal connected the upper and lower segments of the airway posteriorly in the midline. In a study of 5,964 pregnancies by McIntosh *et al.* (1954), this anomaly was not seen.

Holinger *et al.* (1954) described a four-week-old infant who required an emergency tracheostomy for the management of severe respiratory distress and who then failed to respond to resuscitation.

Necropsy showed a cartilaginous obstruction of the cricoid, leaving a small posterior opening.

Smith and Bain (1965) reported nine cases and reviewed the other cases published up till then. They made a classification of this malformation and distinguished three types, corresponding to three different stages in the embryological development of the larynx.

Fox and Cocker (1964) concluded that, in combination with congenital cricoid stenosis, other congenital malformations are frequently found.

In 1968 McMillan and Duvall gave a review of the literature on this

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subject and concluded that the anomaly occurs more frequently than was thought.

Pathogenesis

In order to understand how congenital stenoses are formed, it is useful to recall some well-known embryological facts.

The thyroid cartilage is derived from the fourth arch. The intrinsic muscles of the larynx, the cricoid and arytenoid cartilages and the vocal cords are derivatives of the sixth arch.

Thus a cricoid stenosis will be the result of a defect in the development of the sixth arch.

According to Frazer (1953), the upper end of the laryngotracheal tube becomes obliterated by active proliferation of entodermal cells in an embryo of approximately seven weeks and 5 mm in length. This results in a fusion of the epithelium. At nine weeks, recanalization occurs and the developing larynx becomes larger and more oval-shaped. According to this theory, a cartilaginous stenosis is the result of recanalization after the appearance of chondrification centres in the cricoid cartilage. Therefore a congenital cricoid stenosis, as described by France and Stirling (1960), McMillan and Duvall (1968) and Potter (1962) probably represents a persistence of the earlier stages, in which the cartilage shows a bulky appearance (Wind, 1970).

Diagnosis

Shortly after birth the child develops severe respiratory distress.

The airway impairment may be mild and may manifest itself only on exertion or during an upper respiratory tract infection (Holinger, 1961).

If the airway impairment is complete and resuscitative measures (including emergency tracheostomy) have failed, the obstruction will be found during necropsy.

If a tracheostomy has been successfully performed, then direct laryngoscopy and laryngography, in two directions, will confirm the initial diagnosis.

In all three cases described below, the lateral cervical X-ray proved to be of utmost importance.

Case reports

Case 1

A girl, born on December 14, 1970, showed progressive dyspnoea immediately after birth.

On January 13, 1971, an emergency tracheostomy had to be performed.

Direct laryngoscopy on March 5, 1971, revealed normal, adducted vocal cords, but apparently the subglottic space was occluded by cartilaginous tissue.

A hair-like canal in the posterior commissure made the passage of a 1 mm probe possible. This was also described by Potter (1962).

Laryngography showed the subglottic obstruction, with the hair-like canal at the posterior aspect of the larynx (Fig. 1). No other congenital malformations were found. On March 15, 1971, the cartilaginous obstruction was removed at

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direct laryngoscopy with a biopsy forceps. The child was then intubated nasotracheally with a siliconized PVC tube Ch.16, as reported previously by Berkovits (1971); Bos *et al.* (1973) and Berkovits *et al.* (1974).

The tube was changed weekly and the subglottic space was injected with hyaluronidase 300 U and methylprednisolone acetate 40 mg. This was done three times (every three weeks). After being intubated for half a year the child

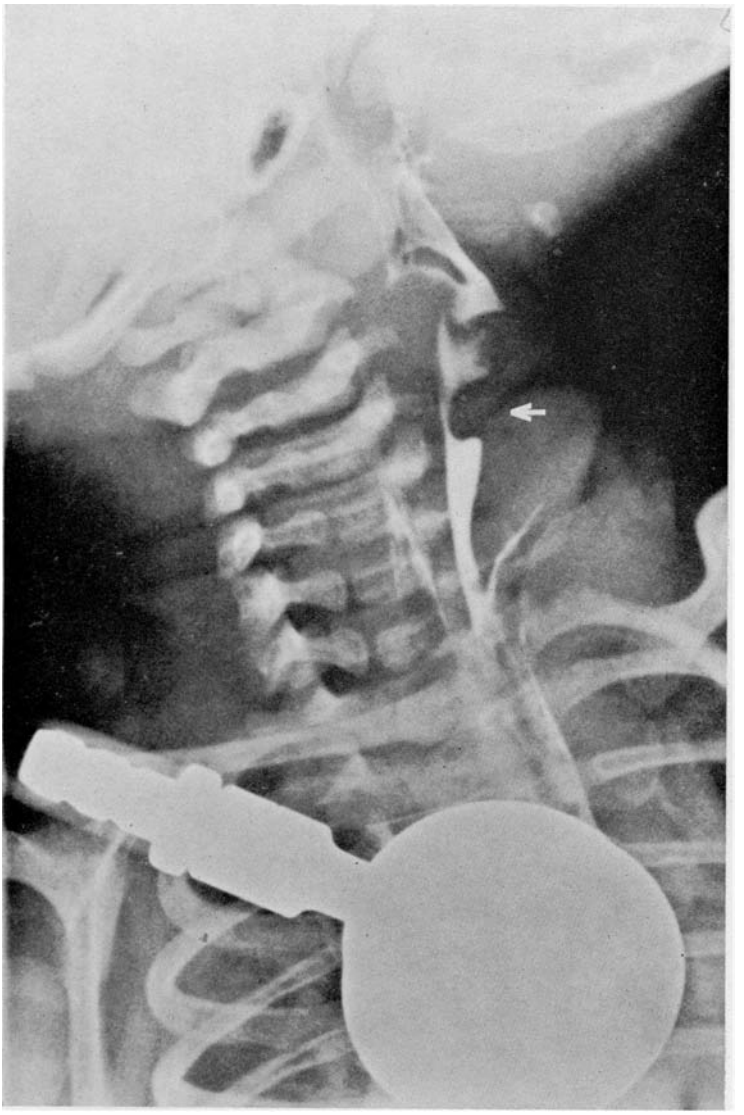


FIG. 1

A laryngogram of case 1.
The subglottic obstruction with a hair-like canal at the posterior aspect of the larynx is marked with an arrow.

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could finally be extubated on September 15, 1971, and her respiration has remained unimpaired to date.

A lateral cervical X-ray made six months after the extubation showed an adequate airway (Fig. 2), and laryngography 6½ years later showed an unobstructed airway (Fig. 3).

As the mother was anxious about the child's speech development, the Foniatric Department was consulted in July, 1973. Their assessment of the girl's

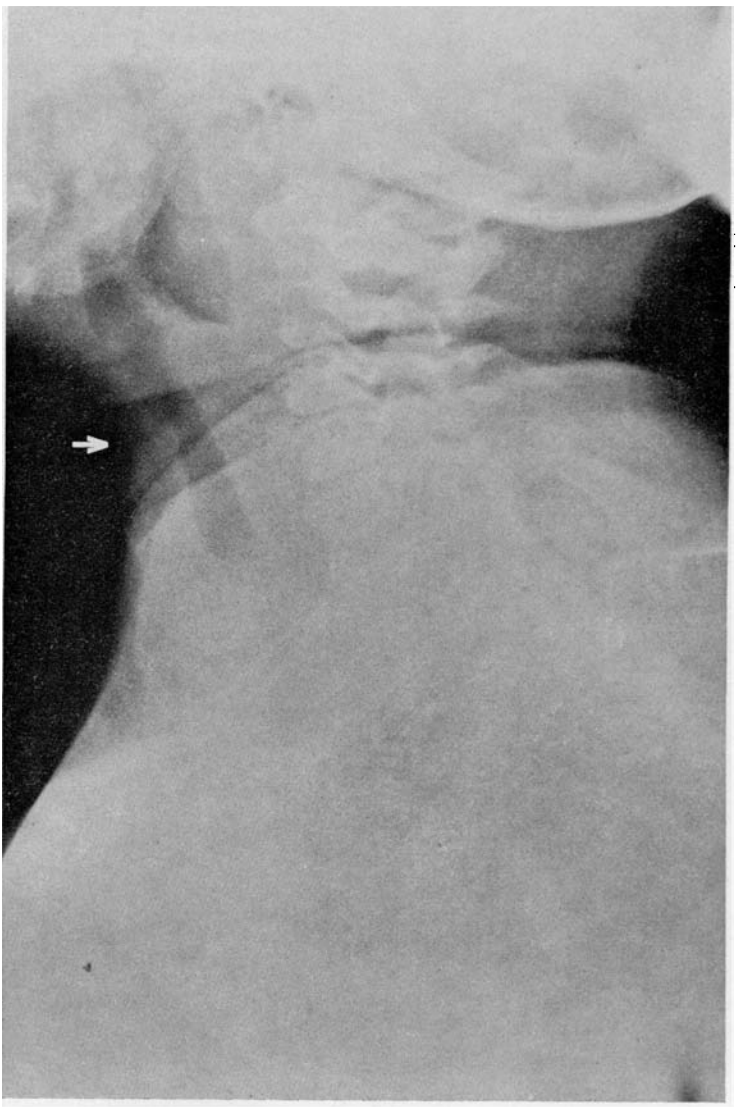


FIG. 2

A lateral cervical X-ray of case 1, made six months after extubation and showing an adequate airway. The arrow marks the site of the former obstruction.

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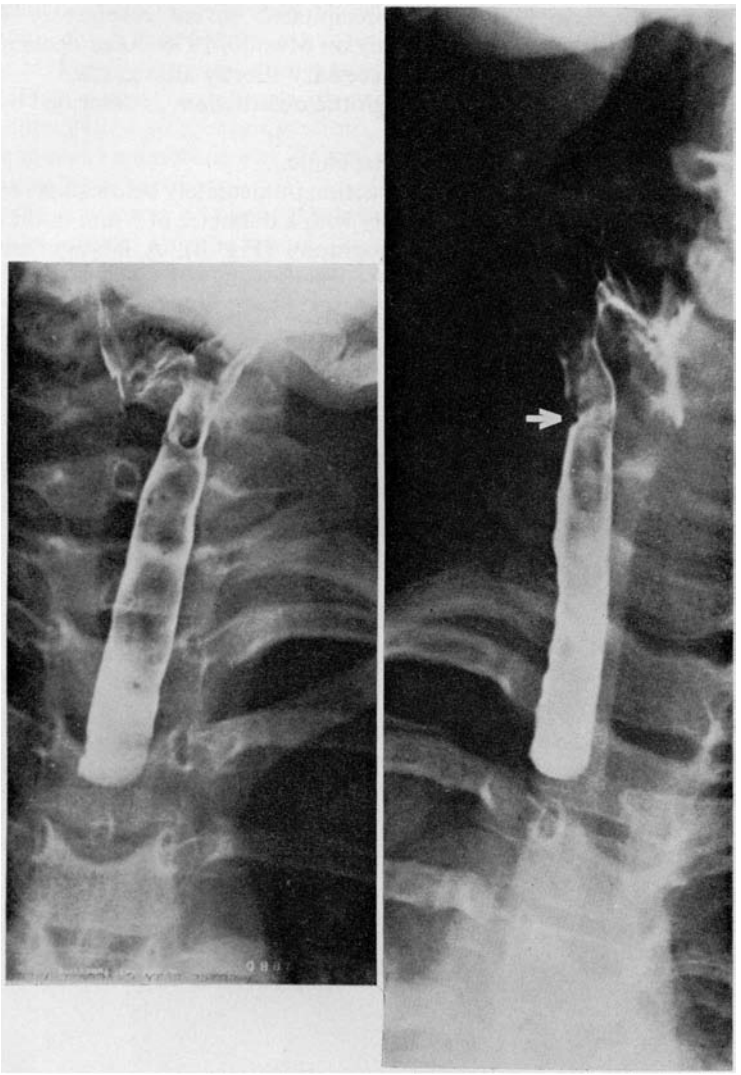


FIG. 3

A laryngogram of case 1, 6½ years after treatment.
There is an unobstructed airway. The arrow marks the site of the former obstruction.

voice revealed that her speech was normal for her age and, consequently, logopedic treatment was not considered necessary.

Case 2

A boy, born on May 8, 1974, was admitted to an Italian hospital in June 1976 with a congenital subglottic stenosis. No other malformations were found.

The child had had an audible respiration since birth, occasionally of a stridorous character.

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An acute respiratory infection precipitated severe respiratory distress, necessitating an emergency tracheostomy on May 20, 1976. After decannulation a second tracheostomy proved to be necessary shortly afterwards.

Tomography revealed a typical subglottic obstruction, as described by Potter (1962).

This patient was then referred to our clinic.

At direct laryngoscopy a large obstruction immediately below the vocal cords was found, leaving an airway posteriorly with a diameter of 3 mm in the sagittal plane. This was confirmed by laryngography (Fig. 4). A laryngofissure was

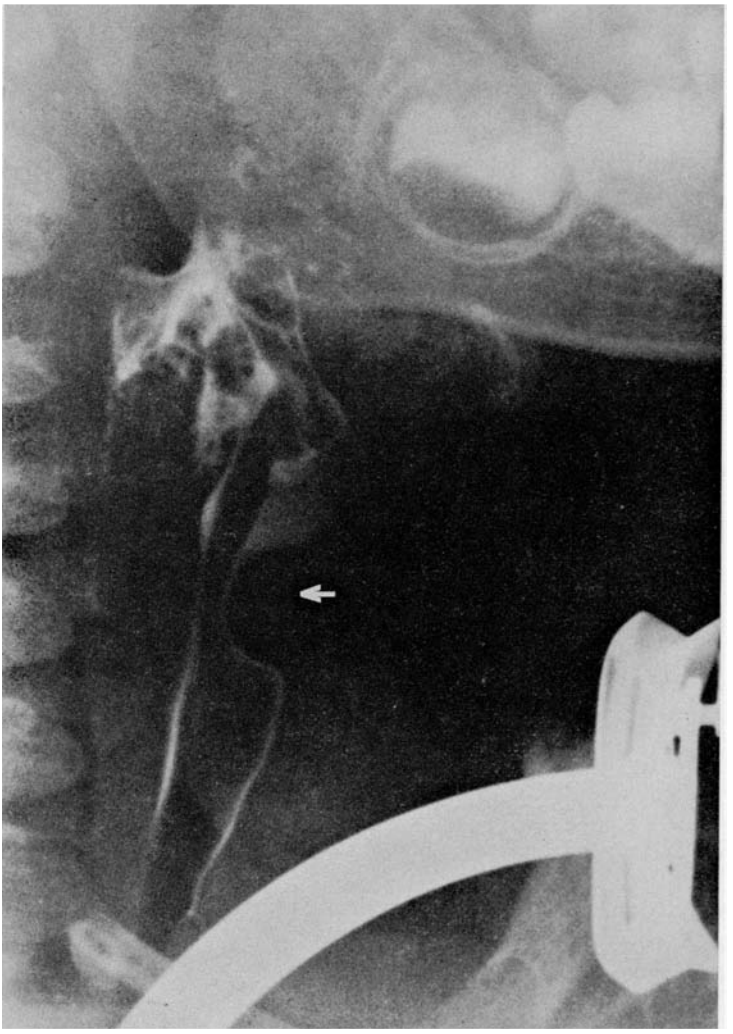


FIG. 4

A laryngogram of case 2.
The subglottic obstruction with a hair-like canal is marked with an arrow.

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performed on June 23, 1976 (Fig. 5), when the cricoid cartilage was found to be solid, with only a hair-like canal posteriorly.

The cartilaginous obstruction was removed with a specially designed curved gouge (Figs. 6 and 7), designed by one of us (R.N.P.B.), using an operating microscope with a 10× magnification.

The mucous membrane was preserved as far as possible and then replaced on the cartilaginous surfaces.

Finally a mixture of methylprednisolone acetate 40 mg and hyaluronidase 300 U was injected into the lesion. The tracheostomy cannula was removed and a nasotracheal tube 18 Fr introduced. This tube was siliconized, according to our previously published method (Berkovits, 1971; Bos *et al.*, 1973).

After weekly re-intubation, the tube was finally removed on July 21, 1976, and recovery then proceeded uneventfully.

On May 5, 1977, direct laryngoscopy and laryngography revealed entirely normal glottic, subglottic and tracheal structures (Fig. 8). The mucous membrane at the site of the former obstruction was smooth and there was no sign of scar tissue formation.

The period of intubation was four weeks and the child has been doing well to date.

Case 3

A boy, born on March 4, 1976, had had an audible respiration since birth, and an acute respiratory infection made it necessary to perform an emergency tracheostomy in August 1976, after attempts at intubation had failed.



FIG. 5

Picture taken during laryngofissure on case 2. A midline incision of the cartilaginous obstruction was made and the cricoid split like an apple. The two arrows mark the obstruction.

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FIG. 6

Picture taken during laryngofissure on case 2. The cartilaginous obstruction of the cricoid, marked with two arrows, has been removed with a specially prepared gouge (Fig. 7).

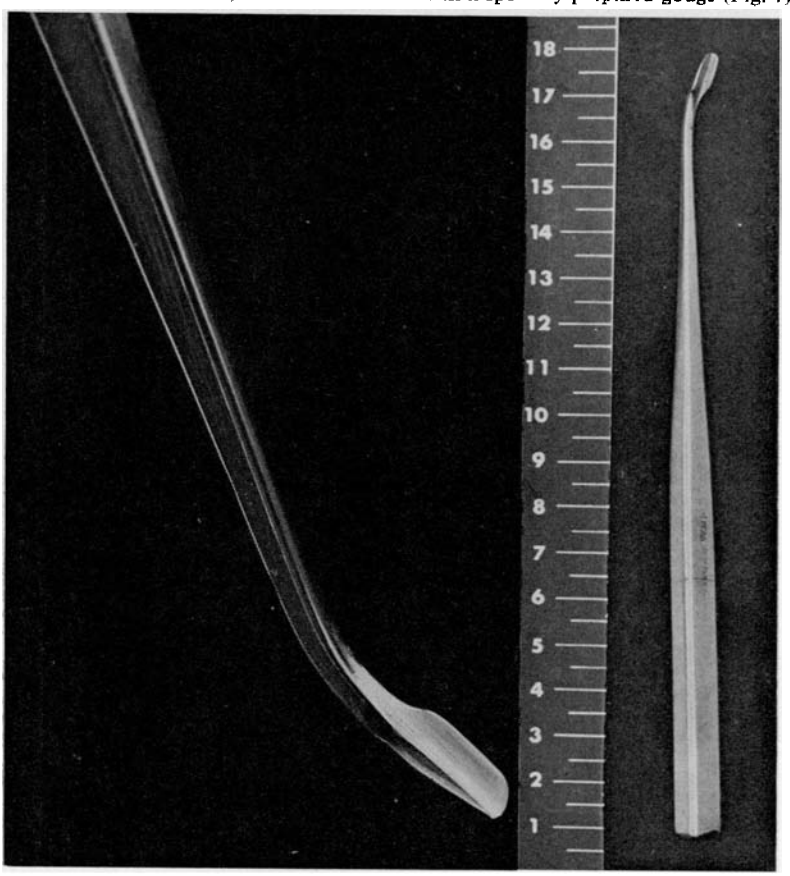


FIG. 7

The specially curved gouge.

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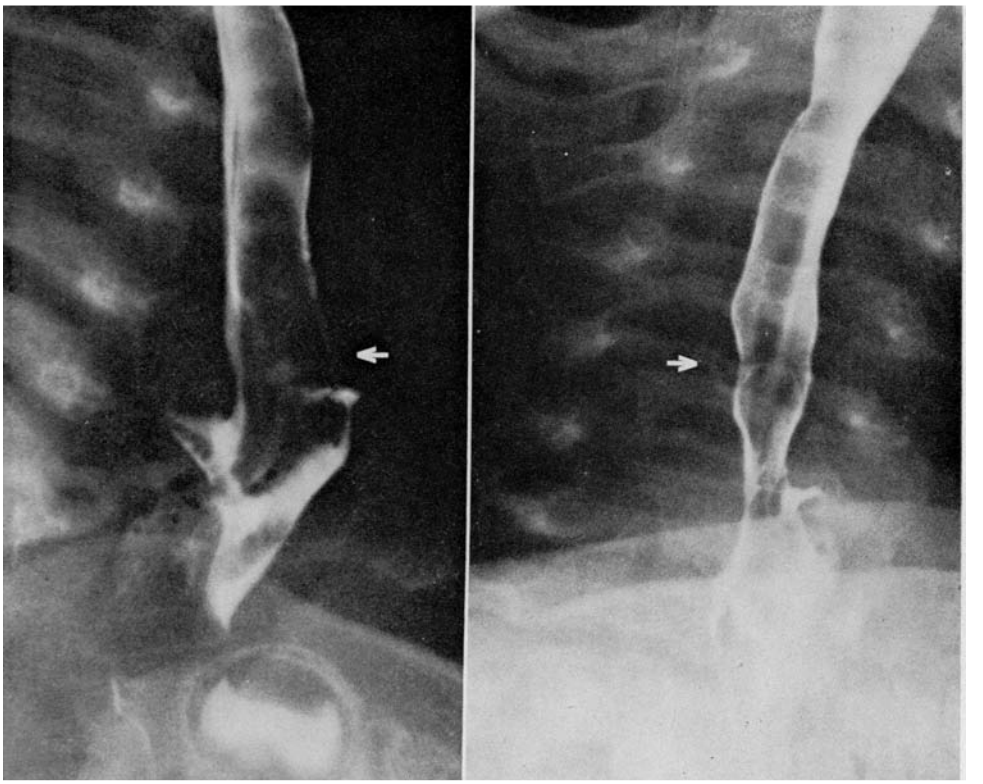


FIG. 8

Lateral and dorso-ventral laryngograms of case 2, made three months after the operation, showing a normal airway. The arrows marks the site of the former obstruction.

Some weeks later, the child was admitted to our hospital. At direct laryngoscopy a *total* obstruction at the level of the vocal cords was found.

Thereafter, laryngography was done, with the child suspended by the feet, and contrast medium was administered through the tracheostomy stoma. This showed an obstruction at the level of the cricoid, with a hair-like canal posteriorly, and complete fusion of the vocal cords (Fig. 9).

On October 15, 1976, a laryngofissure was performed. The cricoid cartilage proved to be solid except for a hair-like canal posteriorly.

Using an operating microscope with 10× magnification, excess cartilage was removed with the curved gouge (Fig. 6). The mucous membrane was preserved as far as possible and then replaced carefully on the cartilaginous surfaces. At direct laryngoscopy the fused vocal cords were cut in the midline. Methylprednisolone acetate 40 mg and hyaluronidase 300 U were injected into the lesion and, finally, a siliconized silicon rubber tube 14 Fr was introduced.* The child was extubated on December 20, 1976. During the period of intubation, the tube

* This silicone rubber tube was kindly supplied by Mr. Jansen, Searle, Uden, The Netherlands.

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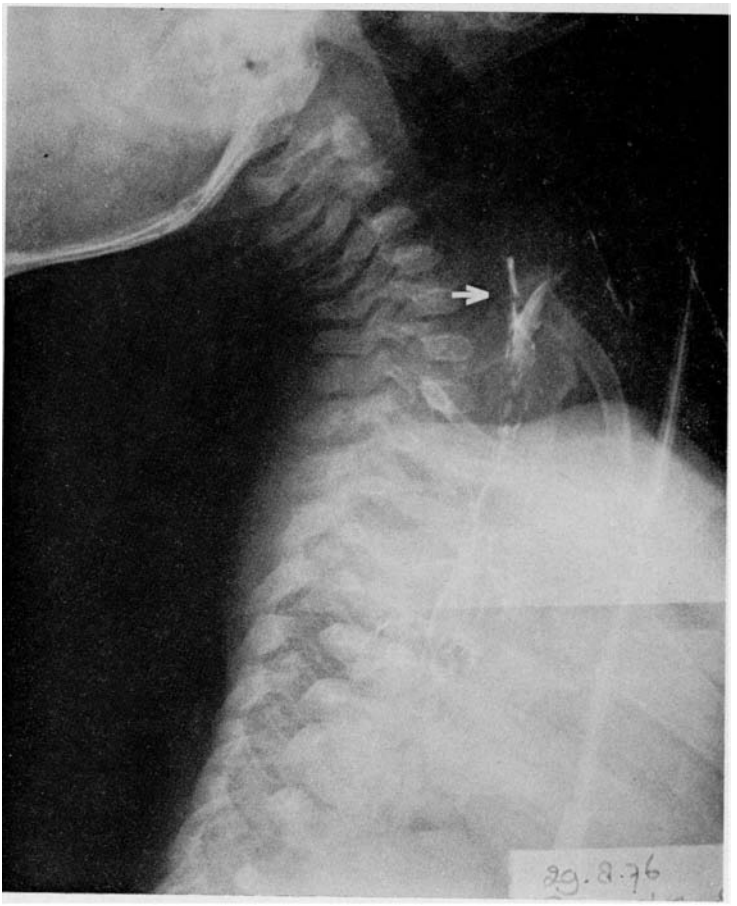


FIG. 9

A lateral laryngogram of case 3, made with the patient suspended by his feet. The hair-like canal is marked with an arrow. The vocal cords are completely fused.

had been changed every two weeks and methylprednisolone acetate 40 mg and hyaluronidase 300 U had been injected into the lesion several times.

Direct laryngoscopy and laryngography, done on January 19, 1977, revealed an adequate airway (Fig. 10). Respiration was unimpaired and patient was discharged from hospital on January 20, 1977.

The period of intubation was eight weeks.

The child has remained well to date. His respiration is unimpaired, but his voice is slightly hoarse.

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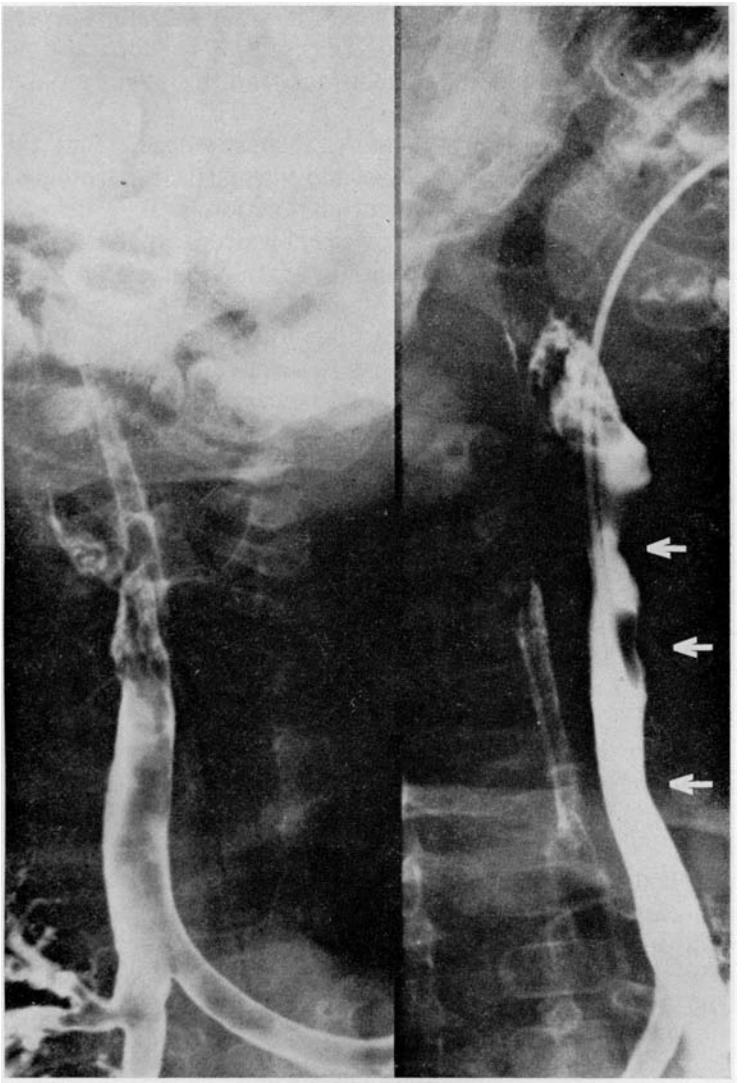


FIG. 10

Lateral and dorso-ventral laryngograms of case 3, made three months after the operation, showing an adequate airway. The upper arrow marks the site of the former congenital obstruction. The middle arrow marks the site of the former tracheostomy. The lower arrow marks a slight narrowing of the trachea at the level of the tip of the tracheostomy cannula.

Discussion

The literature reviewed shows that, in cases of severe subglottic stenosis, an effective method of treatment has yet to be achieved.

In our first case, the obstruction was removed at direct laryngoscopy, because we were afraid that the laryngeal development would be adversely

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affected by a laryngofissure. The effect of laryngofissure on the developing canine larynx was studied by Calcaterra *et al.* (1974). One must indeed use caution when drawing clinical conclusions from results of animal experiments.

However, the results of Calcaterra's study suggest that the infant larynx may be carefully divided by a mid-line vertical thyrotomy without impairment of laryngeal development or function.

In our second and third patients, we performed laryngofissures, with the intention of reducing the long period of intubation of our first patient which had been 26 weeks.

After the laryngofissure, the cartilaginous obstruction was removed under an operating microscope with a 10× magnification. In this way, all the remaining mucous membrane could be carefully preserved and excess cartilage could be removed with the utmost precision. After replacing the mucous membrane on the cartilaginous surfaces and removing the tracheostomy cannula, nasotracheal intubation was performed with specially prepared tubes. In the first two patients we used siliconized PVC tubes (Bos *et al.*, 1973). In the third patient, however, we used (for the first time) an improved model of a silicone rubber tube impregnated with silicone oil 20 cS. This oil is supposed to provide lubrication, thus reducing the friction between the tube wall and the larynx (see also Appendix). (Details of the biological properties of this new low-friction tube will be reported later.)

In this way the properties of 'boundary' lubrication were combined with long-term 'weeping' lubrication. In the last fairly desperate case, we thus achieved a cure after a comparatively short intubation-period of eight weeks.

Our operative technique and subsequent method of treatment have only been carried out on three patients. However, the results of clinical assessment, supported by radiological findings, appear to justify our belief that our method of treatment offers a further advance in the treatment of severe cricoid stenosis.

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R. N. P. Berkovits,
Department of Oto-, Rhino-, Laryngology,
(Prof. Dr. E. H. Huizing),
University Hospital Rotterdam,
Rotterdam, The Netherlands.

A. W. J. de Gee (Appendix),
Metal Research Institute T.N.O.,
Apeldoorn, The Netherlands.

APPENDIX

Friction measurements on silicone tubes—preliminary experiments

In therapeutic laryngo-tracheal intubation, movement of the tube can cause irritation of the mucous membrane. In this respect, it may be expected that the magnitude of the friction force between tube and larynx is important. Indeed clinical assessments of the intubations, described in this paper, strongly suggest that impregnation of silicone rubber tubes with a friction-reducing silicone oil of 20 cS is beneficial.

In order to quantify this friction-reducing effect, with the objective of opening up possibilities for rapid screening of different materials and oils, preliminary experiments of the 'pin-on-disc' type were performed with non-impregnated silicone rubber as well as with silicone rubber, impregnated with silicone (dimethylpolysiloxane) oils, with viscosities of 20 cS and 350 cS respectively.

Equipment

The essential components of the equipment for friction measurement are shown in Fig. A1. An overall view is presented in Fig. A2.

A pin (A, Fig. A1) can be pressed against a disc B, which is mounted on a turntable. The pin is mounted in a specimen holder (C), attached to a thin vertical shaft (E) via a horizontal beam (D). The shaft (E) can rotate with very little friction in two pivot bearings (F). The centre line of the shaft (E) coincides with the centre line of the shaft of the turntable. The rotational movement of the pin holder (C) is restricted by means of a thin wire,

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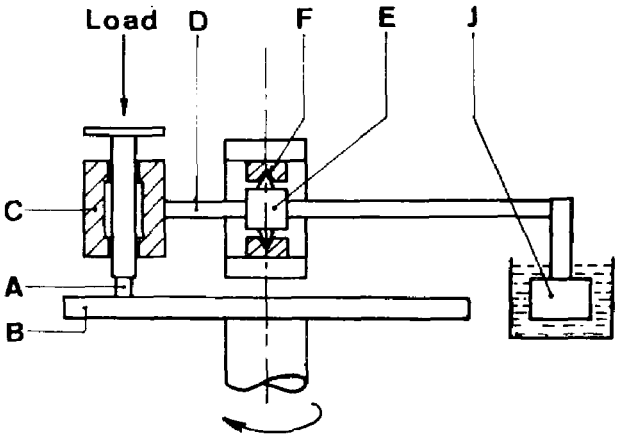


FIG. A1
Schematic presentation of the essential parts of the pin-and-disc test equipment.

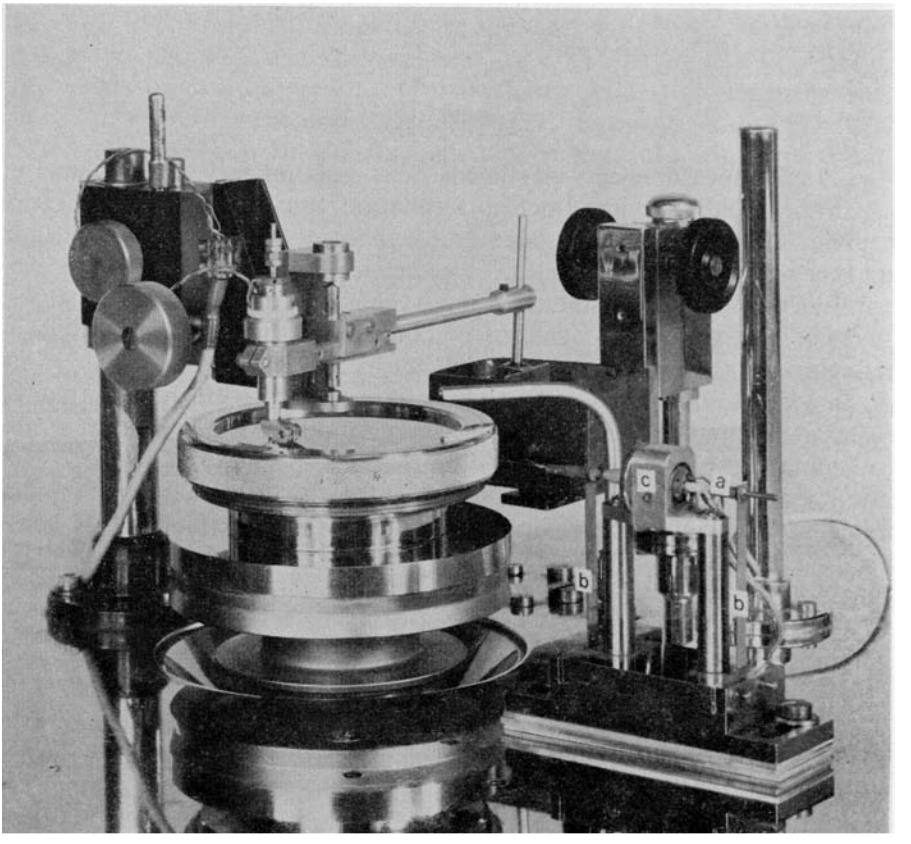


FIG. A2
Overall view of test equipment.

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attached to (C) and connected to a horizontal rod (a), suspended between two vertical blade springs (b) (see Fig. A2).

Friction between pin and disc results in a small deflection of the blade springs and thus in a small displacement of (a). The magnitude of this displacement and, thereby, that of the frictional force between pin and disc, can be measured with an inductive transformer (A). Vibrations in the friction measurement system are damped out with an oil damper (J), attached to the outer end of beam (D).

The pin can be loaded by dead weights, the accuracy of the loading system being about 0·01 N.* During a test, the decrease in height of the pin, due to wear, can be measured continuously with an inductive transducer (K). The temperature of turntable and disc can be kept constant within 1 °C by pumping water of constant temperature through channels, machined in the turntable. The whole assembly is surrounded by a glove box, into which gas (e.g. air) of carefully controlled humidity can be introduced.

As can be seen in Fig. A2, the disc is mounted on the turntable by means of an overlapping screw ring. The shallow chamber—thus formed—can be filled with liquid. In one series of tests (see below) it was filled with distilled water of 21 °C.

Materials

In the present preliminary tests a disc of stainless steel 316 was used. Prior to application, this was given a mirror polish. The pins were made of hemispherically tipped rods of silicone rubber of the following dimensions (prior to impregnation; see below): length, 8 mm; radius cylindrical part, 4 mm; radius tip, 2 mm. The cylindrical part was completely surrounded by a close-fitting aluminium socket. Fig. A3, left, shows a non-impregnated pin, ready for use.

Gravimetric analysis showed that—upon impregnation—silicone rubber absorbs 40–50 wt per cent of silicone oil of 20 cS viscosity.

As shown in Fig. A3, middle, this causes considerable swelling of the material. Of 350 cS oil only 15–20 wt per cent is absorbed; consequently less swelling occurs (Fig. A3, right).

Experimental conditions

The experiments were performed under the following conditions:

normal force on the pin	: 0·10 N
speed of rotation of the disc	: 10 rev./min
linear speed of the sliding	: 0·02 m/s
experiment duration	: ½ h
temperature	: 21 °C
environment	: air of 60 per cent relative humidity and distilled water under air cover

* 1 N = 1 Newton ≈ 102 gram force (gf).

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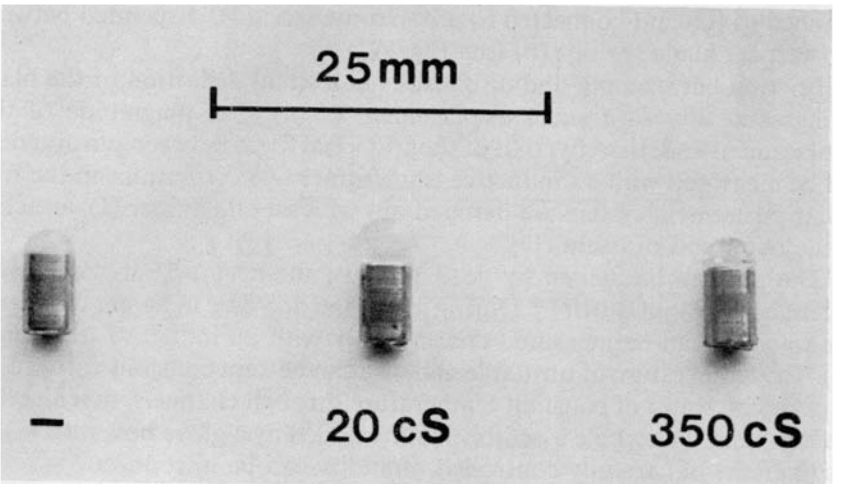


FIG. A3

Pins of silicone rubber, prior to testing.
Left : non-impregnated.
Middle: impregnated with 20 cS oil.
Right : impregnated with 350 cS oil.

Results and discussion

In Figs. A4 and A5, results are presented in the form of reproductions of recorder outputs, showing friction force (from left to right) as a function of time (from bottom to top). Fig. A4 shows the results of tests performed with a dry disc. It can be seen that, with a non-impregnated pin, the friction force fluctuates considerably, i.e. from about 0·13 N to about 0·18 N (thus—at a normal force of 0·10 N—the coefficient of friction varies between 1·3 and 1·8). The frequency of this fluctuation is 10 min $^{-1}$, i.e. it is coupled directly to the speed of rotation of the disc. It is probably caused by the occurrence of minor irregularities in the topography of the disc surface.

Fig. A4, middle, shows that impregnation with silicone oil of 20 cS leads to an appreciable reduction in the amplitude of fluctuation in the friction force. Also, the maximum value of the friction force is reduced considerably, i.e. after 15 min of sliding $F_{max\,(15)}$ from about 0·18 N to about 0·08 N.

Finally Fig. A4, right, shows that impregnation with silicone oil of 350 cS has much less effect upon friction. Actually in this case $F_{max\,(15)}$ reduces to about 0·16 N.

In Fig. A5 the results of the experiments performed in the presence of distilled water are presented. A comparison between Fig. A4 and Fig. A5 shows that in all cases water clearly provides lubrication. Actually, for the non-impregnated pins, $F_{max\,(15)}$ is reduced from about 0·18 N in the dry condition to about 0·08 N in the presence of water. It can also be seen that,

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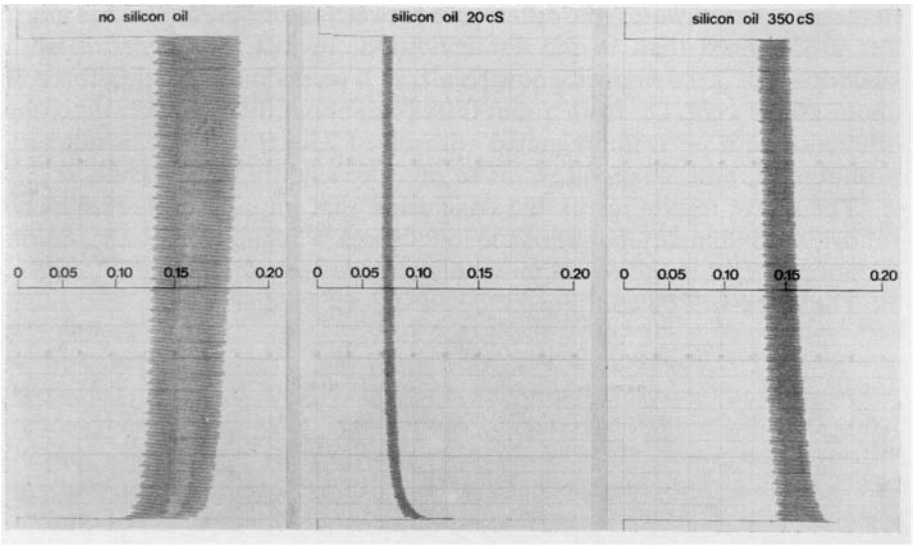


FIG. A4

Reproductions of recorder outputs.
From left to right : friction force in N.
From bottom to top: time.
Environment : air of 60 per cent relative humidity.

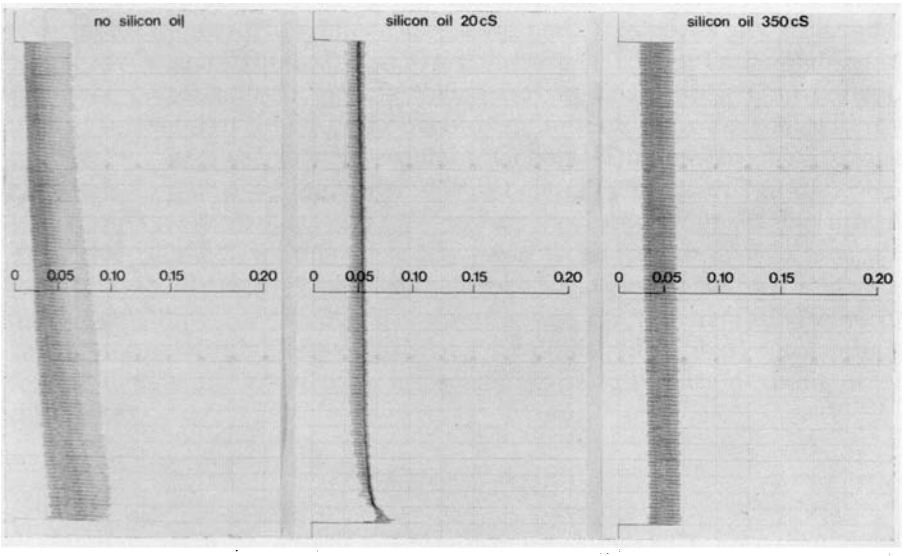


FIG. A5

Reproductions of recorder outputs.
From left to right : friction force in N.
From bottom to top: time.
Environment : distilled water of 21 °C.

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in the presence of water, the differences between the different pins are much less pronounced than in the dry condition. In fact, impregnation with silicone oil of 20 cS viscosity now results in a reduction of friction force of about 25 per cent, i.e. from about 0·08 N to about 0·06 N. This time, the difference with pins, impregnated with oil of 350 cS viscosity, is not very pronounced. Still, 20 cS oil seems to produce slightly better results.

The above results justify the conclusion that impregnation of silicone rubber with dimethylpolysiloxane oil causes a significant reduction in friction, at least in combination with stainless steel.

The work will be continued.

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