# Brief Clinical Report

# Retrograde fibreoptic intubation in a child with Nager's syndrome

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***Purpose:*** *The authors describe a retrograde fibreoptic technique for tracheal intubation in a micrognathic child with a tracheo-cutaneous fistula.*

***Clinical features:*** *A four-year-old child with Nager's syndrome presented for surgical closure of a tracheocutaneous fistula. A tracheostomy tube had been placed in the neonatal period for management of upper airway obstruction due to severe micrognathia. At 2½ yr of age, after a successful mandibular advancement procedure, the tracheostomy was removed and the child allowed to breathe through the natural airway. Preoperative physical examination revealed an uncooperative child, unable to open her mouth due to limited temporo-mandibular motion. The child was first anaesthetized with ketamine, 70 mg im, then halothane by mask. The authors were unable to open the child's mouth sufficiently to allow rigid laryngoscopy. Attempts at oral and nasal fibreoptic intubation were unsuccessful. Ultimately, the authors were able to intubate nasally by passing an ultrathin Olympus LF-P laryngoscope under direct vision through the tracheocutaneous fistula in a cephalad direction, through the larynx and nasopharynx, then out the nares. An endotracheal tube was then advanced over the fibreoptic scope and positioned distal to the tracheocutaneous fistula. The surgical procedure was successfully accomplished and the trachea was extubated postoperatively without difficulty.*

***Conclusion:*** *Retrograde fibreoptic intubation may be an option for airway management of a select group of children who cannot be intubated by traditional techniques.*

***Objectif:*** *Décrire la technique d'intubation endotrachéale fibroscopique utilisée par voie rétrograde chez un enfant micrognathe et porteur d'une fistule trachéocutanée.*

***Caractéristiques cliniques:*** *Un enfant de quatre ans porteur du syndrome de Nager était programmé pour la fermeture chirurgicale d'une fistule trachéocutanée. Préalablement, à la période néonatale, une canule de trachéostomie avait été insérée pour traiter l'obstruction des voies aétiennes causée par une importante micrognathie. À deux ans et demi, après une intervention réussie pour avancer la mandibule, la trachéostomie avait été délaissée pour permettre à l'enfant de respirer par les voies naturelles. L'examen préopératoire montrait un enfant non coopératif et incapable d'ouvrir la bouche suffisamment pour permettre la laryngoscopie rigide. L'enfant a d'abord été anesthésié à la kétamine 70 mg im et ensuite au masque avec halothane. Les tentatives d'intubation par laryngofibroscopie orale et nasale se sont avérées infructueuses. Finalement, les auteurs ont réussi à intuber pr voie nasale en passant un laryngoscope Olympus LF-P ultrafin sous vision directe à travers la fistule trachéocutanée en direction céphalique, à travers la larynx et le nasopharynx et ensuite à travers une narine. Le tube endotrachéal a été ensuite poussé sur le fibroscope et positionné distalement à la fistule trachéocutanée. L'intervention chirurgicale a pu être effectuée et la trachée extubée en postopératoire sans difficultés.*

***Conclusion:*** *L'intubation fibroscopîque rétrograde peut représenter une solution applicable à la gestion des voies aériennes chez un groupe choisi d'enfants qui ne peuvent être intubés par les techniques conventionnelles.*

**Key words**

ANAESTHESIA, paediatric: Nager's syndrome;
INTUBATION: fibreoptic, retrograde;
SYNDROMES: Nager's.

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*Accepted for publication 15th March, 1996.*

Many techniques have described the establishment of the airway in children whose tracheas are known to be difficult to intubate by rigid laryngoscopy due to severe micrognathia.[1] When rigid laryngoscopy is impossible, either a retrograde technique or oral/nasal fibreoptic laryngoscopy may facilitate intubation.[1–3] We describe airway management in a child with Nager's syndrome, whose trachea was unable to be intubated by both rigid laryngoscopy and nasal/oral fibreoptic laryngoscopy. A

retrograde fibreoptic intubation technique for airway management is described.

## Case report

This four-year-old 12.3 kg child with Nager's syndrome presented for tracheocutaneous fistula closure and temporo-mandibular joint examination under anaesthesia. Nager's syndrome was diagnosed shortly after birth, manifested by typical craniofacial abnormalities (including severe micrognáthia), cleft palate, and characteristic limb deformities. An emergency tracheostomy was performed under local anaesthesia shortly after birth after multiple attempts at awake intubation by rigid laryngoscopy failed. Subsequent anaesthetics, including two for mandibular lengthening procedures, were managed without difficulty with ventilation provided via the existing tracheostomy. At age 2½ yr, otolaryngological reevaluation demonstrated a patent upper airway; the tracheostomy tube was removed. With the exception of extremely limited mouth opening and a tracheocutaneous fistula, the child was doing well at home.

Preoperative physical examination revealed a small, uncooperative child with limited mandibular mobility. An air leak was present at the site of the previous tracheostomy. Preoperative routine laboratory data were normal. Anaesthesia was induced with 70 mg ketamine *im* with 0.25 mg atropine. Mask oxygen was applied and ventilation assisted without difficulty. One percent halothane was added to the inspired gases as intravenous access was being obtained. After ensuring that the tracheocutaneous fistula would allow placement of a 3.0 mm endotracheal tube, if necessary, the fistula was manually occluded to prevent air leakage during assisted ventilation.

Pancuronium 1.2 mg *iv* was administered and ventilation controlled. Even after adequate muscle relaxation, the mouth would not open wider than two centimetres, preventing successful placement of a rigid laryngoscope. Fibreoptic intubation attempts both orally and nasally were unsuccessful. Mask ventilation was continued without difficulty. Ultimately, an ultrathin LF-P fibreoptic intubating laryngoscope (Olympus Corp., Lake Success N.Y.) was placed through the tracheocutaneous fistula, directed cephalad through the trachea, through the larynx, and into the nasopharynx. Under direct visualization, the fibreoptic scope was advanced out of the nares, and a 4.0 mm endotracheal tube threaded over the fibreoptic scope. The endotracheal tube was fully advanced into the trachea, the fibreoptic scope removed, and the endotracheal tube positioned immediately distal to the tracheocutaneous fistula without difficulty (Figure 1). Following surgical closure of the tracheocutaneous fistula, neuromuscular blockade was antagonized with neostigmine 0.8 mg and atropine 0.4 mg *iv*, the child awakened, and the trachea was extubated without difficulty.

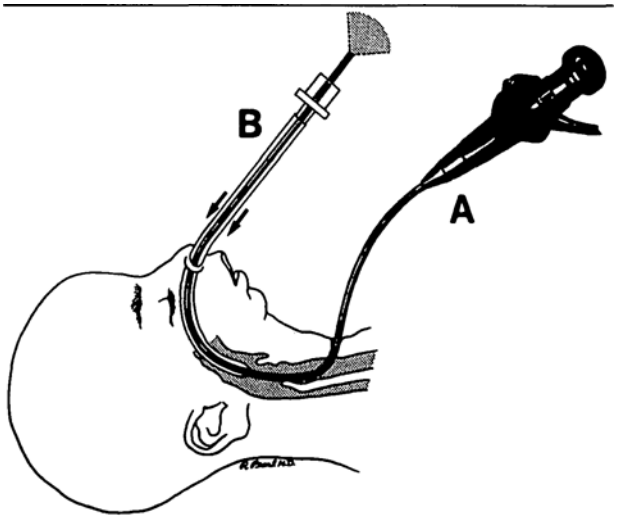


FIGURE 1 Retrograde fibreoptic intubation: (A) The LF-P fibreoptic scope is placed through the tracheocutaneous fistula, into the trachea in a cephalad direction, through the vocal cords, through the nasopharynx, exiting the nares. (B) The endotracheal tube is threaded over the fibreoptic scope and positioned in the mid trachea, then the fibreoptic scope is with-drawn.

## Discussion

Nager's syndrome is a rare form of acrofacial dysostosis, which includes characteristic craniofacial abnormalities (Figure 2).[4–6] As in the related Miller's and Treacher-Collins syndromes, airway management in Nager's syndrome may be extremely difficult.[4–8] Upper airway obstruction secondary to severe micrognathia often necessitates tracheostomy placement in many of these children.[4]

Children with Nager's syndrome require multiple surgical interventions in early life for correction of orthopaedic and craniofacial abnormalities. Inhalational mask anaesthesia may be unsuccessful due to the severe nature of the upper airway deformity. Awake oral intubation would seem rarely indicated, with no guarantee of success. Historically, failing rigid laryngoscopy, children with severe micrognathia have been intubated using a retrograde intubation technique.[1,3] Currently, these children's tracheas can be intubated either after sedation or induction of inhalational anaesthesia with the use of an oral or nasal fibreoptic technique.[1,2,9]

The child we report posed an unique challenge. Closure of the tracheocutaneous fistula required that the airway be secured from above the old stoma site. Ankylosis of the temporo-mandibular joints made direct

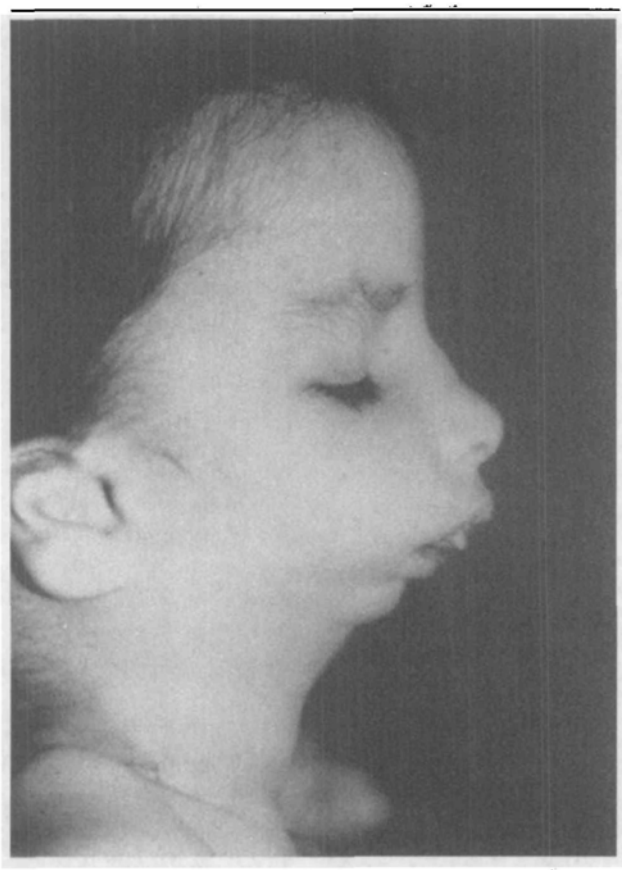

FIGURE 2 Typical craniofacial features of Nager's syndrome include downward slanting palpebral fissures, malar hypoplasia, ear deformities, and severe micrognathia.

laryngoscopy impossible, prompting our use of the flexible fibreoptic intubating scope. The LF-P intubating scope, with external diameter of 2.2 mm., allows placement through even the small diameter endotracheal tubes used in infant anaesthesia.[10] The child's previous mandibular lengthening procedures resulted in a patent upper airway, allowing good spontaneous ventilation after intramuscular ketamine. As inhalational agents were added, ventilation was assisted without difficulty. Our inability to intubate the larynx fibreoptically from above was due to a combination of secretions obscuring the fibreoptic visual field (no suction port is available in the LF-P fibreoptic scope) and the distortion of the child's airway anatomy. The trachea was ultimately intubated using a retrograde technique. We passed the ultrathin scope under direct visualization in a cephalad direction, through the tracheocutaneous fistula, past the vocal cords, into the nasopharynx, and exiting the nares. It was then a relatively simple manoeuvre to pass the endotracheal tube over the fibreoptic scope (using it as a stylet) and thread the endotracheal tube into the trachea.

The circumstances we have described in the care of this child are not likely to occur frequently in anaesthetic practice. Nevertheless, use of an ultrathin fibreoptic laryngoscope in a retrograde manner may not be immediately obvious to the practitioner facing a similar circumstance. Retrograde fibreoptic intubation should be added to the list of airway management techniques in children whose trachea cannot be intubated by traditional techniques.

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