

# Mucopolysaccharidosis and Anaesthesia

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# Introduction

- Normal Anaesthesia
- Problems Associated with MPS
- Techniques
- Safety

# Normal Anaesthesia

- Pre-operative assessment/ preparation
- Induction of Anaesthesia
- Maintenance of Anaesthesia
- Emergence
- Analgesia

# Airway

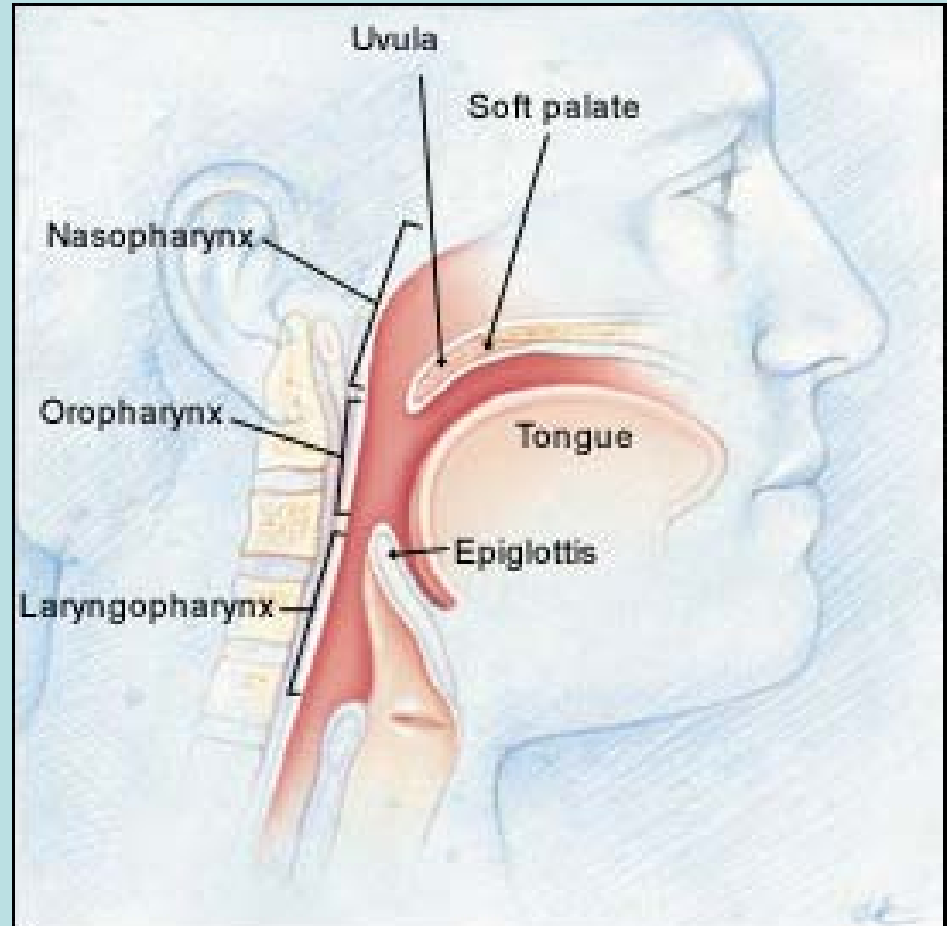


Illustration © 1999 Chréty Krames

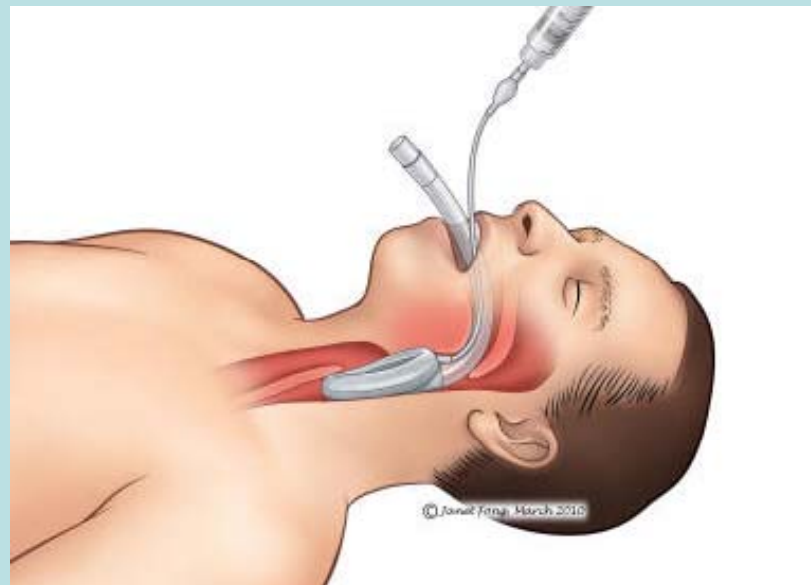
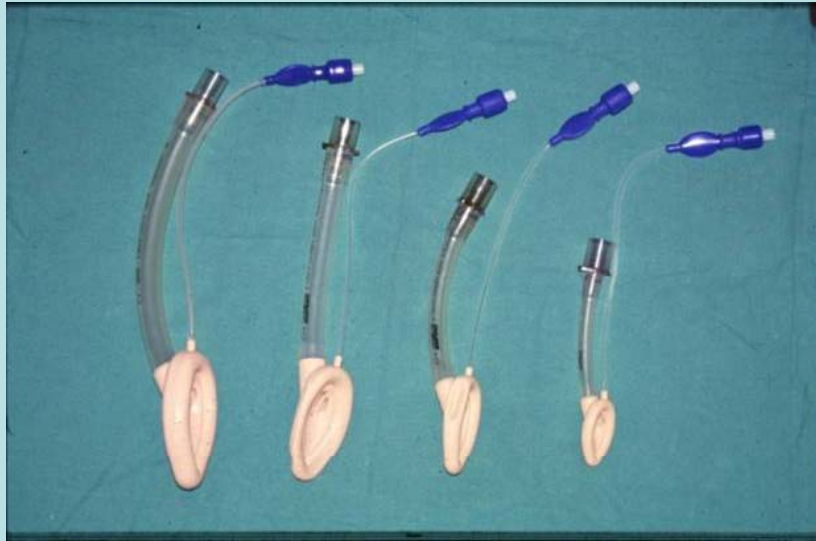
# Airway Maintenance



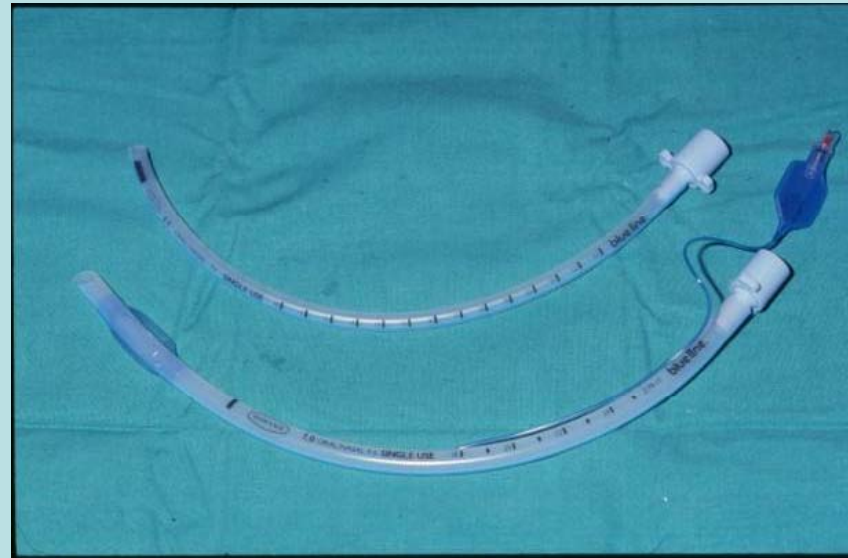
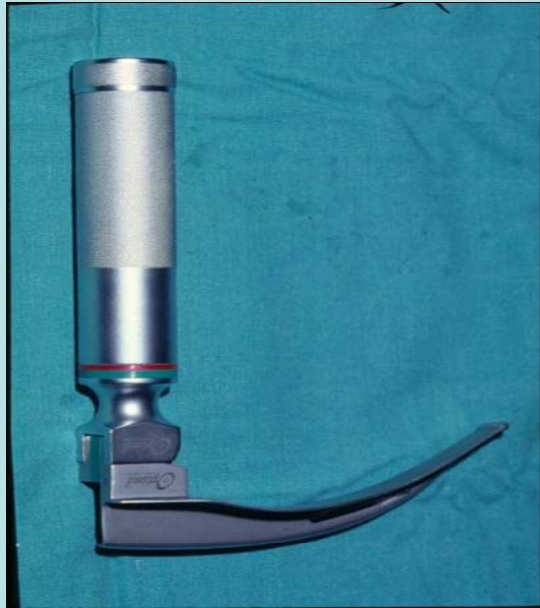
# Airway Adjuncts



# Laryngeal Mask



# Endotracheal Tube





# Normal Intubation



# The MPS Anaesthetic Issues

- Poor Cooperation
- Difficult iv access
- Difficult Airway
- Difficult Intubation
- Cervical Spine Instability
- Respiratory Disease
- Cardiovascular Disease
- Care needed with positioning

# Team Approach

- Physician
- Surgeon
- Anaesthetist
- ENT specialist
- Theatre staff
- Intensive care staff

# Pre-operative preparation

- ECG, Echocardiogram
- Chest X ray
- Blood gases
- Oxygen saturation (overnight)
- Naso–endoscopy
- Anaesthetic visit
- Note review

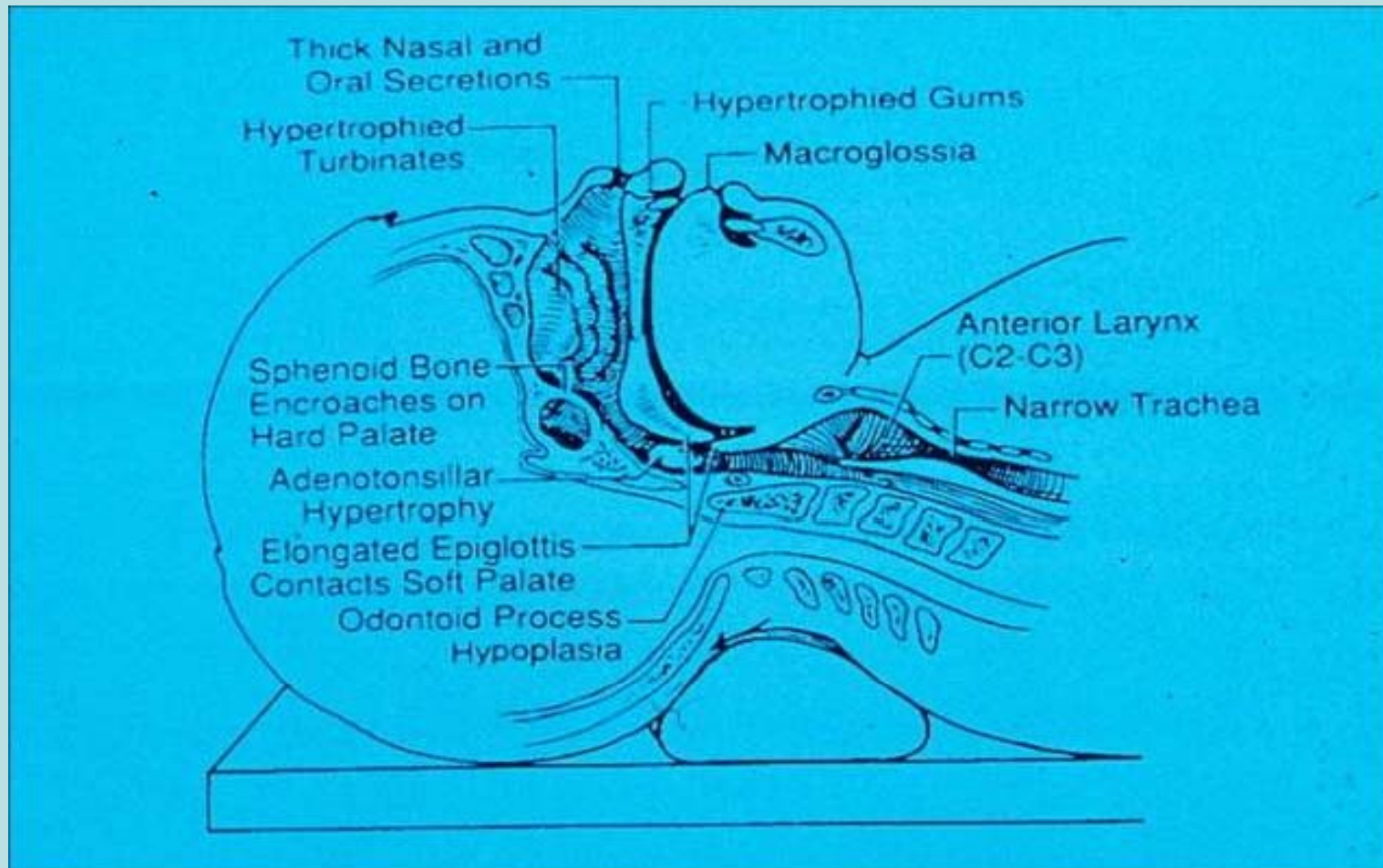
# Pre-medication

- Reduce anxiety
- Dry secretions
- Provide Analgesia
- Support the cardiovascular system
- Commence anaesthesia
- **BALANCE OF RISKS**

# The MPS Airway



# MPS Airway



# Induction

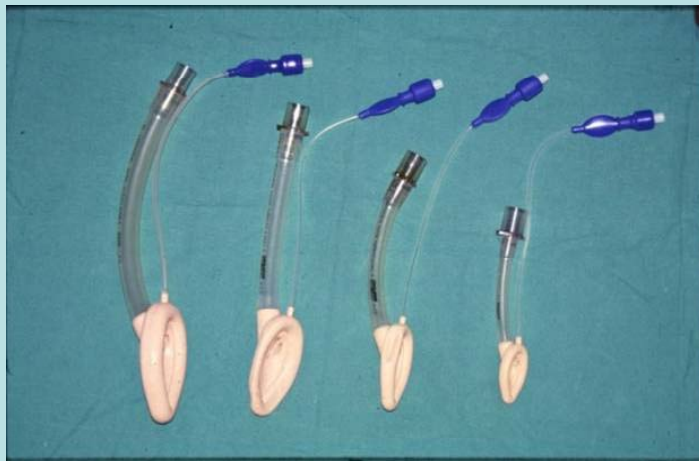
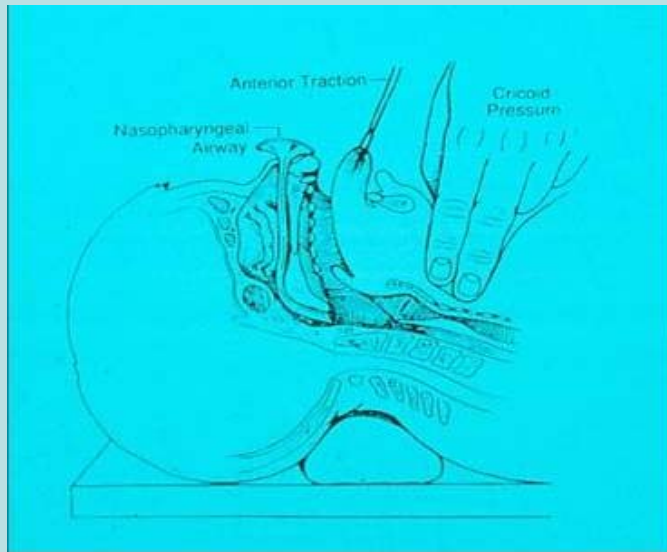
- Intravenous
- Gaseous
- Awake intubation
  
- **PRINCIPLE IS TO MAINTAIN  
SPONTANEOUS RESPIRATION**



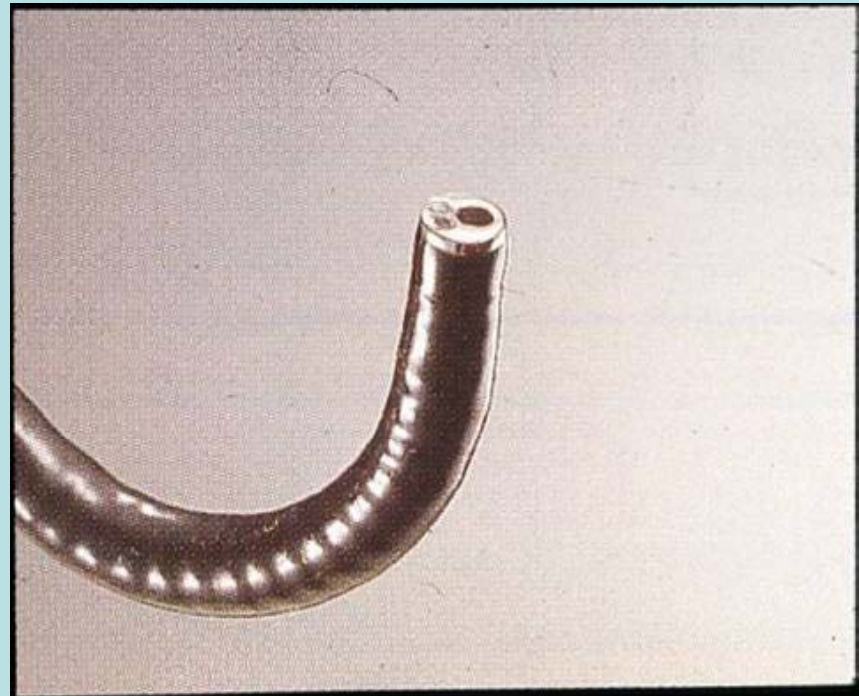
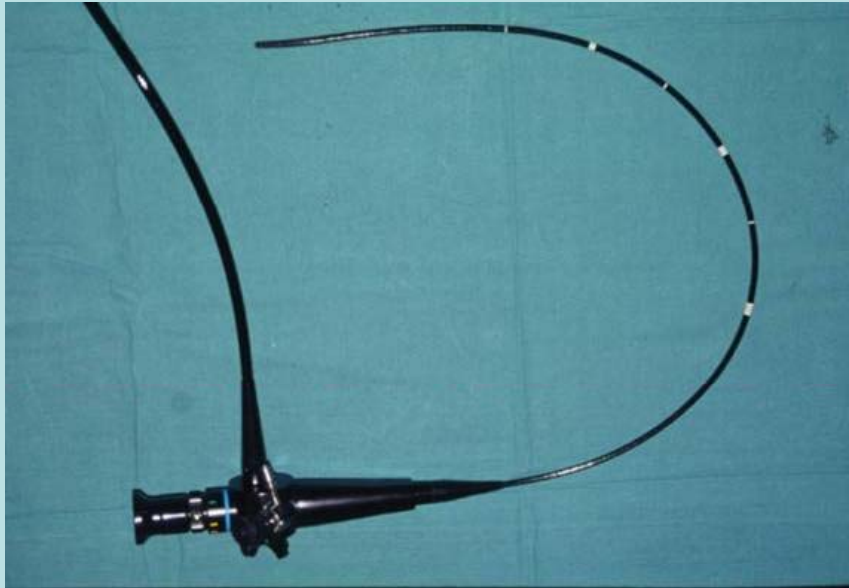
# Gaseous Induction



# Airway Manoeuvres/ adjuncts

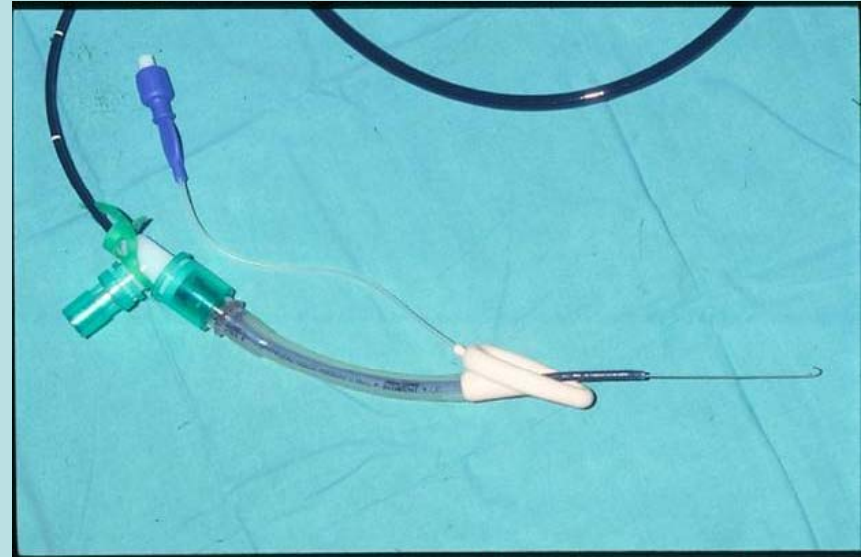


# Fibrescope

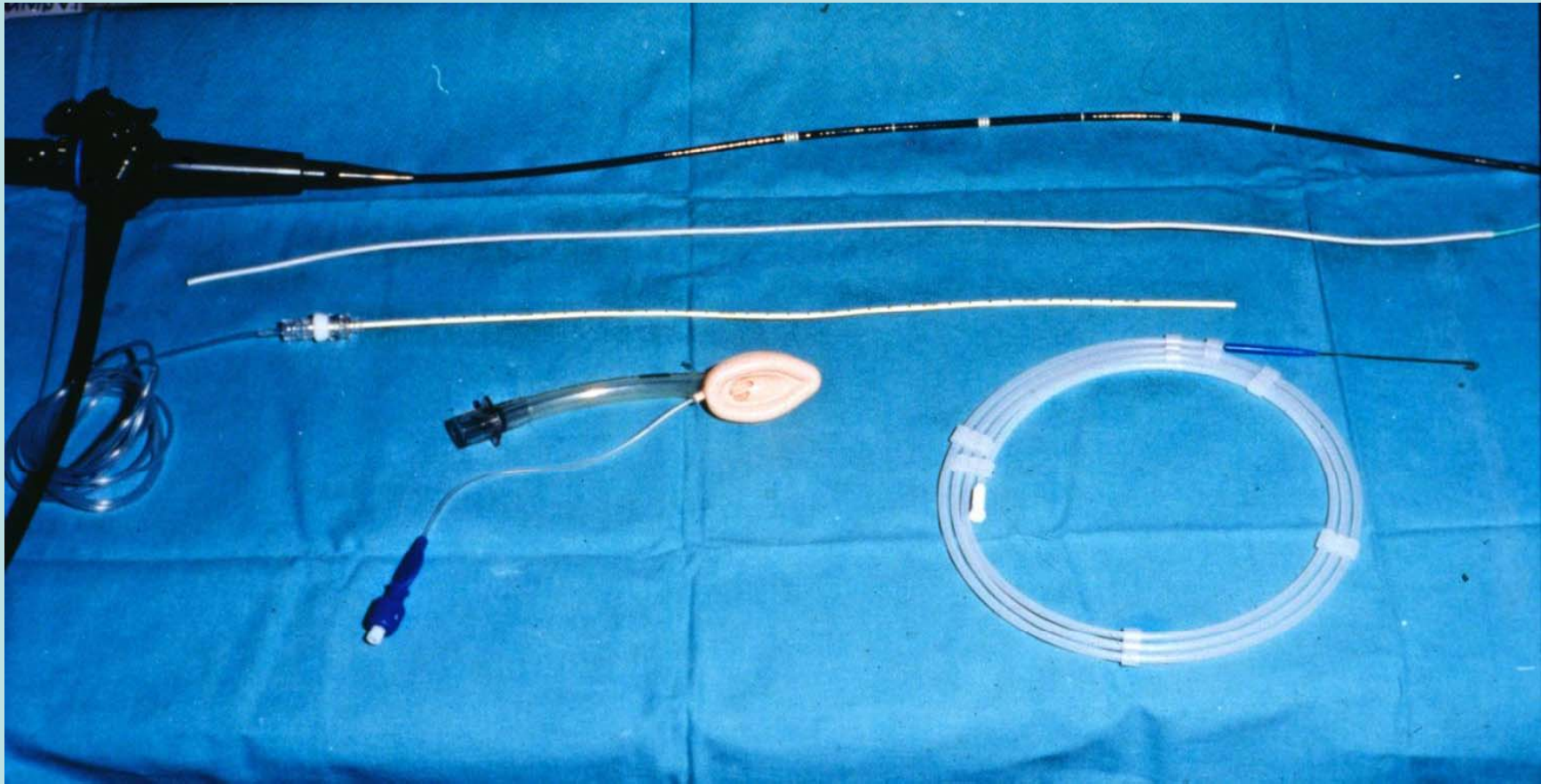


# Fibreoptic Intubation

- Nasally
- Orally
- Orally via an Adjunct
  - LMA



# Equipment





**How safe is an MPS  
anaesthetic?**

*Anaesthesia*, 1994, Volume 49, pages 1078–1084

REVIEW ARTICLE

## **Anaesthesia and mucopolysaccharidoses**

**A review of airway problems in children**

**R. W. M. WALKER, M. DAROWSKI, P. MORRIS AND J. E. WRAITH**



- Royal Manchester Children's Hospital
- 1989-1991
- 34 children
- Mixture of all MPS sub-types
- 89 anaesthetics



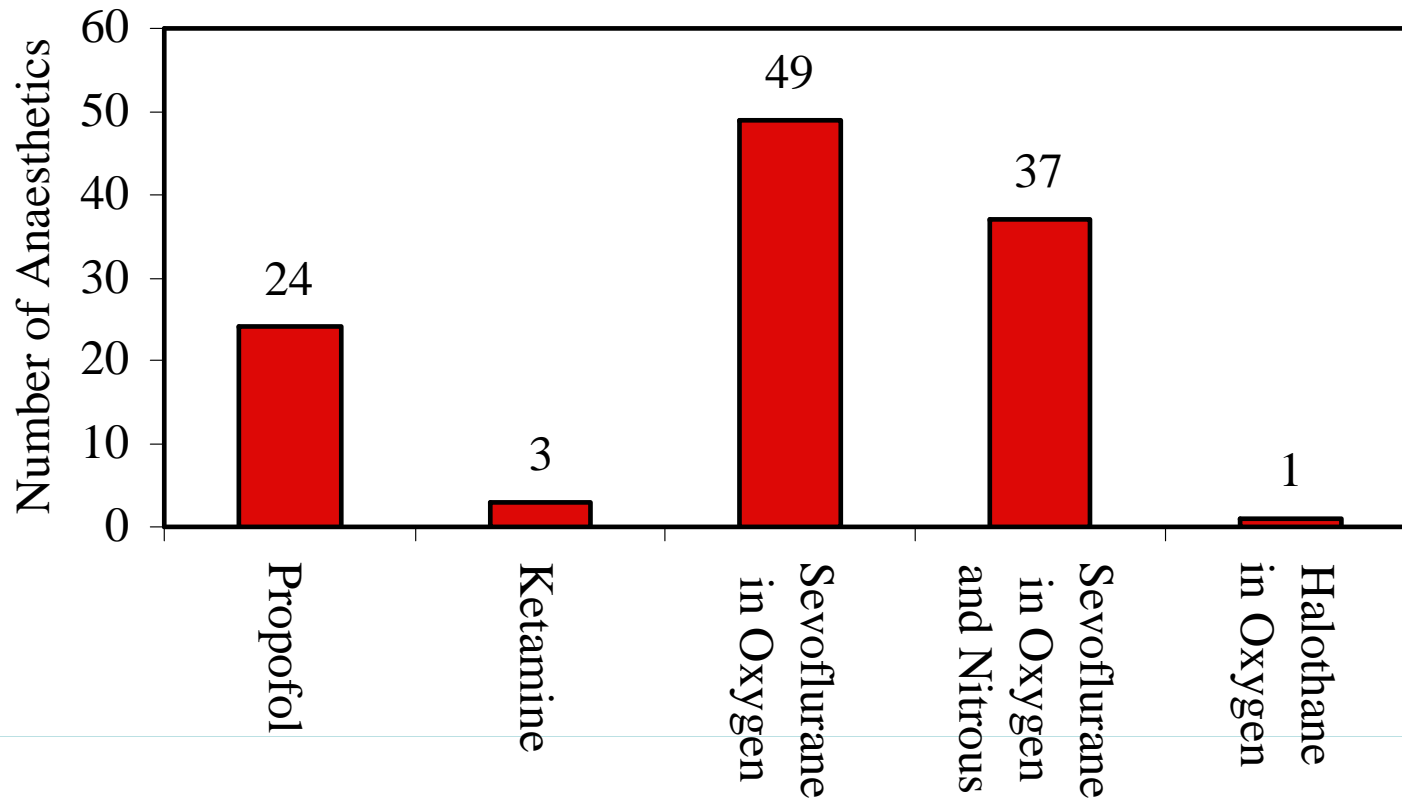
MPS	1H	1H BMT	1HS	II	IV	VI	ML
No patients	7	1	4	7	1	8	1
Age range	1-8	3-4	16-22	9-13	13	2-16	10
No. Intubs	11	2	8	9	1	12	1

- 60 intubation attempts in 29 patients
- 15/29 presented no difficulty
- 11/29 noted to be difficult (38%)
- 3/29 intubation failed (8%)

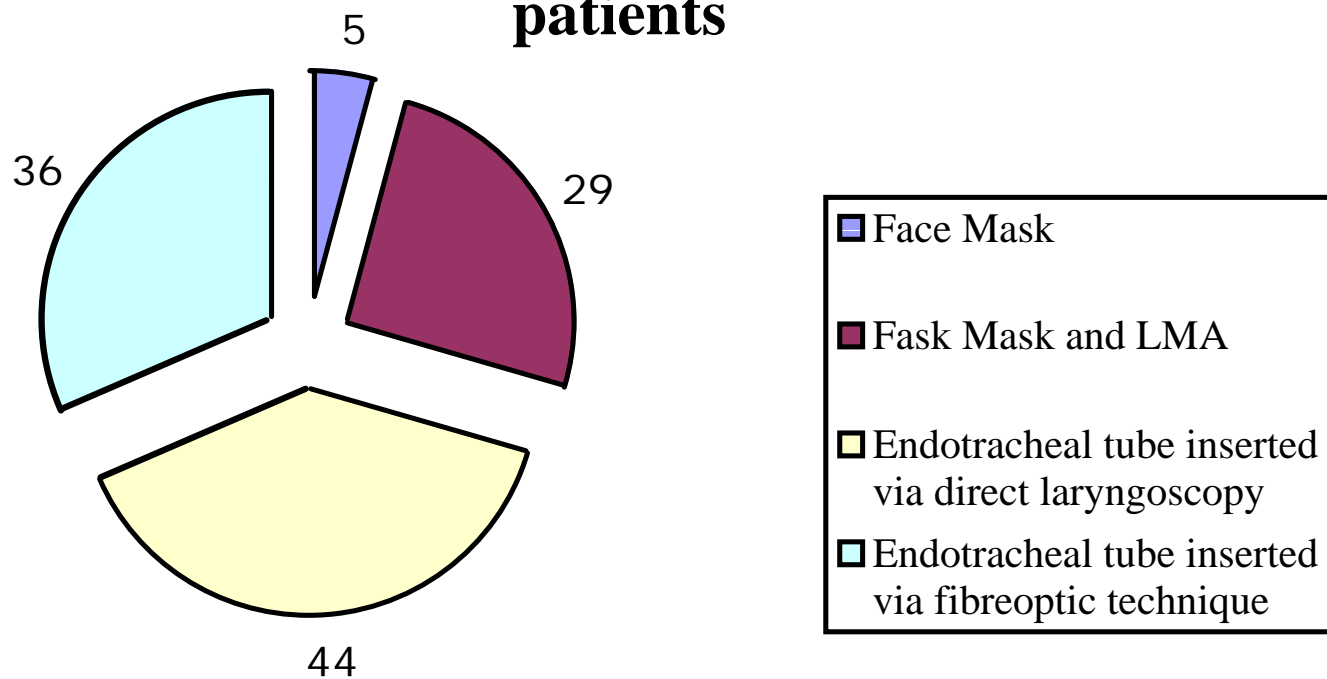
# More Recently

- 2000-2008
- MPS I
- 40 children
- 114 Anaesthetics
- 37/114 on ERT
- 58/114 had BMT
- 2/114 ERT and BMT
- 17/114 Neither

## Induction techniques used for patients with MPS I



## Airway devices used for Anaesthesia in MPS patients



# Airway

- 18/114 peri-operative problems
- 8 instances in 6 patients with either airway obstruction or difficult bag ventilation on induction
- All resolved by either airway adjunct or turning lateral

# Intubation

- Intubation attempted 80/114
- 44/80 Direct laryngoscopy
- 36/80 Fibre-optic intubation via LMA
- 6 instances in 5/40 patients of significant issues in securing the airway
- 2 due to MPS deposits in airway (procedure abandoned)
- 1 unsuccessful (procedure continued on LMA)

# Post-op

- 3 instances in 2/40 patients of airway obstruction in recovery
- 101/114 patients went directly to the ward
- 13/114 went to critical care
- 2/13 intubated (1 planned)



# Summary

- Anaesthesia in MPS is not as hazardous as it once was and can be achieved relatively safely in most MPS patients
- The risks associated are much reduced as a result of
  - Advances in Medical Treatment
  - Greater Awareness and training of Anaesthetists
  - Technology
- MPS patients will present in greater numbers to the adult anaesthetist

# Thankyou

