

The impact of contemporary treatments on the perioperative care of children with mucopolysaccharidoses: A case series and review of the literature.

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Abstract

Background: Patients with mucopolysaccharidosis (MPS) present significant perioperative challenges. We aimed to document the perioperative care of children with MPS undergoing anesthesia and to describe the impact of hematopoietic stem cell transplantation (HSCT) or enzyme replacement therapy (ERT) on the anesthetic management of these patients.

Methods: We performed a retrospective chart review of patients with MPS anesthetised for surgical or investigative procedures at the Hospital for Sick Children in Toronto between January 2000 and December 2014. Data on MPS treatment, co-morbidities, anesthetic techniques, airway management, post-operative care and perioperative complications were collected.

Results: 66 children with MPS underwent 332 anesthetics for 345 procedures. The overall rate of difficult airway was 42%. Of 29 patients with MPS I (Hurler syndrome), 66% were treated with HSCT and 34% with ERT. In those treated with HSCT, 19% had difficult airways, compared with 67% in patients who received neither treatment. 90% of patients with MPS II (Hunter syndrome) had difficult airways. ERT did not improve airway difficulty in MPS I or II patients. 32% of all anesthetics were conducted without airway instrumentation. Direct laryngoscopy, was used in 26% of all anesthetics, the laryngeal mask airway in 26%, fiberoptic bronchoscope in 7%, and video laryngoscope in 5%.

Conclusions: Patients with MPS I who have had HSCT are less likely to have a difficult airway compared with those not treated with HSCT. ERT in MPS I and II patients did not alter the incidence of difficult airway. A third of MPS patients underwent anesthesia for diagnostic imaging or minor interventional radiology procedures without airway instrumentation.

Background

The mucopolysaccharidoses (MPS) are a related group of inherited lysosomal storage disorders involving inappropriate cellular accumulation of glycosaminoglycans, resulting in progressive multisystem complications including organo-megaly, airway abnormalities, cardiovascular and respiratory disease, joint and bony deformities, visual, auditory and intellectual deficits, and dysmorphic features.¹⁻³ Classification and characteristics of the different MPS are summarised in Table 1.

Historically the management of patients with MPS has been palliative with a focus on the control of symptoms.¹⁻⁵ Modern treatment options include haematopoietic stem cell transplantation (HSCT), first reported in 1981, and enzyme replacement therapy (ERT) with recombinant alpha-L-iduronidase (Laronidase[®]) being used to treat MPS I since 2004.⁶ Other currently available ERT include: recombinant iduronate-2-sulphatase (Elaprase[®]) for MPS II, recombinant N-acetylgalactosamine 6-sulphatase for MPS IV and recombinant N-Acetylglucosamine 4-sulphatase (Naglazyme[®]) for MPS VI. Both forms of treatment have been shown to reduce the severity of disease progression, however, skeletal and ocular and neurological outcomes remain poor.⁷

Patients with MPS present significant challenges when undergoing anesthesia, particularly in airway management.^{4,8-15} The incidence of difficult airway in untreated patients with MPS I (Hurler syndrome) has been reported to be 54%.⁹ Kirkpatrick et al and Frawley et al have described improvements in the rate of difficult airway in MPS I patients who have undergone HSCT or ERT.^{16,17} Anesthetic techniques such as the use of total intravenous anesthesia may facilitate minimal airway instrumentation for non-invasive procedures. Also, newer airway management techniques and equipment has broadened the options for the control of these notoriously challenging airways.¹⁶

The purpose of this study is to document the perioperative care of children with MPS undergoing general anesthesia at our institution, and to describe the impact contemporary treatments of MPS on anesthesia management and outcomes.

Methods

Approval from the Research Ethics Board at the Hospital for Sick Children was obtained for a retrospective analysis of the records of all children diagnosed with MPS between January 2000 and December 2014. Records were reviewed to determine patient demographics, classification of MPS, treatment with HSCT or ERT, perioperative airway management and the incidence of difficult airways. A difficult airway was defined as a reported difficulty with bag mask ventilation or reported difficulty of visualising the larynx, multiple attempts at intubation, the need for intubation aids due to a failure of direct laryngoscopy, or failure to intubate the trachea by a consultant anesthetist.¹⁸ Data retrieved are presented descriptively.

Results

Patient demographic profiles, metabolic therapies and comorbidities

Sixty-six patients underwent 332 anesthetics for 345 surgical or diagnostic imaging procedures. All children had a confirmed diagnosis of an MPS. Patient demographics are summarised in Table 2.

Procedures performed

Of the 332 procedures performed (Table 2), 53% were done on an ambulatory basis, 41% were admitted electively and 6% were emergency procedures. The mean duration of anesthetic for all procedures was 98 minutes (median 75 minutes). The majority of anesthetics were administered for diagnostic imaging (28%) or interventional radiology (17%). Otorhinolaryngoscopic and orthopedic surgery comprised 12% and 8% respectively. (Table 3)

Anaesthetic delivery

Eight percent (n = 28) of children received sedative premedication, including oral midazolam (75%, n = 21), intravenous midazolam (18%, n = 5) and lorazepam (7%, n = 2). No complications were reported in association with preoperative sedation. 28% (n = 92) of children had intravenous access established prior to induction of anesthesia.

Fifty-seven percent (n = 189) of anesthetics were induced with inhalational agents, including sevoflurane (59%, 196), nitrous oxide (56%, 142), halothane (2%, 5) and isoflurane (5%, 5). 11% (n = 35) had simultaneous intravenous and inhalational induction. Thirty-three percent (n = 108) of anesthetics were induced with intravenous agents including thiopental (9%, n = 10), propofol (81%, n = 87), fentanyl (6%, n = 7) and ketamine (1%, n = 4).

Sixty-seven percent (n = 222) of the anesthetics were maintained with inhalational agents (sevoflurane, isoflurane, desflurane or halothane with or without nitrous oxide) and 33% (n = 109) were maintained with intravenous anesthesia agents, mainly propofol.

Thirty-nine percent (n = 129) of patients did not receive perioperative fluid therapy, 45% (n = 151) received normal saline or Hartmann's solution and 11% (n = 35) received a dextrose containing solution. One patient required blood transfusion.

Airway management

Airway management and incidence of difficult airway is presented in Table 4. The overall rate of difficult airway was 42% (n = 42), and the overall failed intubation rate was 2% (n = 3). 32% of procedures were completed without airway instrumentation (face mask, nasal prongs or regional only). In these cases, patients remained spontaneously ventilating, and anesthesia was maintained with total intravenous anesthesia (propofol) or inhalational agent via face mask. The majority of these anesthetics were for diagnostic imaging or brief interventional radiology procedures.

Of the 29 patients with MPS I (Hurler's syndrome), 16 were treated with HSCT and 7 with ERT, 3 received both ERT and HSCT, and 3 remained untreated. In those treated with HSCT, 19% had difficult airways, compared with 67% in patients who received neither HSCT nor ERT, representing a 72% reduction in airway difficulty. There was no difference in the difficult airway rate in untreated MPS I patients (67%) versus MPS I patients who received ERT (71%). None of the MPS I patients who received both treatments (ERT prior to HSCT) had difficult airways. In 70% of MPS I children, ERT was commenced over 5 years of age, whereas HSCT occurred under 18 months of age in 89% of cases.

Ninety percent of patients with MPS II (Hunter's syndrome) had difficult airways. ERT was not associated with improvement in airway difficulty (100% vs 80%).

Difficult airway management

A total of 28 patients with difficult airways underwent 142 anaesthetics (Table 5). 29% of these patients underwent anesthesia without airway instrumentation (face mask or nasal prongs). The laryngeal mask airway (LMA) was utilised in 23% of their anesthetics and the remaining 47% of cases were intubated. In this group of patients with difficult airways, direct laryngoscopy was used only 8% of the time. Direct laryngoscopy with assistance occurred in 13% of cases. Assisted direct laryngoscopy included the use of backward, upward and rightward pressure on the larynx, a bougie and/or the two-person technique where one experienced anesthetist performs laryngoscopy and manipulates the larynx externally to achieve the best possible view whilst another passes the endotracheal tube through the vocal cords. The fiberoptic bronchoscope was used 15% of the time. 27% of fiberoptic intubations occurred using the LMA as a conduit for the bronchoscope. Videolaryngoscopy was used 11% of the time. Videolaryngoscopy was introduced to our institution in 2006. Of note, its use increased proportionately more in the latter years of this study, with 68% of its use being since 2009. Conversely, the number of fiberoptic intubations have declined overtime, with only 32% of fiberoptic intubations occurring after 2009 in this case series.

Postoperative care and perioperative complications

Post-operatively, 45% (n = 150) of children were discharged on the day of surgery, 47% (n = 157) were managed on a general ward, and 8% (n = 25) were managed in a pediatric intensive care unit. The average length of hospital stay was 14 days; however, this was influenced heavily by the cohort of MPS I (Hurler syndrome) patients who were inpatients during their HSCT. Excluding the children with MPS I, the average length of hospital stay was 5 days.

The most frequent intraoperative complications related to the challenging airways so common amongst children with MPS. Of significance, two cases developed laryngospasm (0.6%) and 5 cases whose airways were not instrumented had obstructive breathing (1.5%). Aspiration occurred in one child. Only 2 children received regional anaesthesia without general anaesthesia (spinal anesthetic combined with ilioinguinal block in one and caudal anesthetic in the other). In another case, caudal anesthesia failed and general anaesthesia was induced.

In the postoperative period, complications were similarly minimal with only four children developing airways obstruction requiring intervention in the post anesthetic recovery unit (1.2%). Of note, the most serious morbidity and mortality were as a result of complications HSCT, with two patients developing graft versus host disease. Four children died whilst still inpatients from complications secondary to HSCT (21% of all patients receiving HSCT).

Discussion And Review Of Mps

Patients with MPS have a 25% overall reported rate of difficult airway, and up to 54% and 71% in patients with Hurler's and Hunter's syndromes, respectively.⁹ HSCT and ERT appear to slow the progression of MPS, although the benefits of HSCT in patients over 18 months of age are reported to be minimal.¹⁹⁻²¹ We report a 72% reduction of difficult airway in patients with Hurler's syndrome treated with HSCT. Compared to HSCT, ERT did not improve airway difficulty in any of the MPS types, however the impact of ERT administered in infancy remains unknown. Regardless of airway difficulty, almost a third of MPS patients were managed without airway instrumentation, suggesting that spontaneous ventilation in these patients can be safely used whether with inhalational agents or total intravenous anesthesia for non-invasive or minimally invasive procedures. Where intubation of the difficult airway is required, videolaryngoscopy appears to be succeeding fiberoptic intubations as the first option in securing the airway.

This study has many important limitations. Firstly, it is a retrospective review of data over many years. The reported results are therefore reliant on the accuracy of documentation for example of airway management. However, although documentation of airway management in normal airways may be incomplete, we have found that where difficult airways were encountered, documentation is often very detailed. As MPS as a whole is a relatively uncommon disease, this study is limited by the small numbers of patients available for inclusion. This makes statistical comparisons difficult and less clinically meaningful.

HSCT treatment

HSCT, including bone marrow transplant and umbilical cord blood transplant; is the treatment of choice for the severe form of MPS I (Hurler syndrome) in patients who are less than two years old with minimal neurological involvement. Lysosomal enzymes require a mannose-6-phosphate residue to target the lysosomal compartments within cells. Transplanted stem cells replace the macrophages of donor marrow, which then release normal enzymes into the serum of affected recipients of the HSCT; the residue binds to mannose-6-phosphate receptors on the deficient host cells and is taken up into the cell by this recognition system. Only 1 - 2 % enzyme activity is needed to avert the clinical damage caused by the host's enzyme deficiency, and the engrafted macrophages provide an ongoing source of normal enzyme.²⁰

Successful engraftment improves survival and within a year can reduce or reverse the somatic progression of the disease in MPS I, including cardiac and airways disease, sleep apnoea and obstruction, intellectual and developmental decline, hearing loss, hepatosplenomegaly and joint stiffness. To optimise the benefit of this intervention, HSCT should be performed at an early age before permanent damage is caused by the accumulation of glycosaminoglycans. Perhaps owing to poor tissue perfusion and enzyme penetration, skeletal (spinal), retinal, and cardiac valvular disease are not altered, and progression can be expected.²¹

In the milder attenuated form of MPS I (Scheie syndrome) the risk of HSCT (15–20% morbidity and mortality) outweighs the benefits of treatment; and in MPS II (Hunter) and MPS III (Sanfilippo) where the neurologic deterioration is not halted by the treatment, HSCT is not usually undertaken.²²

Enzyme replacement therapy

Recombinant ERT is now available for MPS I (Hurler), MPS II (Hunter) and MPS VI (Maroteaux-Lamy). ERT is licenced for use in the United States and European Union; Laronidase (Aldurazyme) for MPS I (Hurler), Idursulfase (Elaprase) for MPS II (Hunter) and Galsulfase (Naglazyme) for MPS VI (Maroteaux-Lamy).²³

Recombinant ERT is administered as a weekly infusion, and has proved to be safe, with minor allergic responses to the infusion being the main side effects. It is indicated as the primary treatment in attenuated MPS I (Scheie syndrome), MPS II (Hunter) and MPS VI (Maroteaux-Lamy), or as an adjuvant to HSCT where it has been shown that administration prior to and after HSCT can improve engraftment.^{21,24}

Clinical improvement after the use of ERT in MPS I has been demonstrated with a 5.6% increase in pulmonary function, improved left ventricular function (although valve disease remains unchanged), increased functional exercise capacity and activities of daily living. It is suggested that earlier commencement of ERT further improve outcomes. ERT does not cross the blood brain barrier and therefore does not improve neurological outcomes.²¹

Current focus of research includes ERT for MPS III (Sanfilippo) and MPS IV (Morquio) as well as the potential of intrathecal administration of these drugs to improve neurological outcomes.

Anaesthetic considerations in the Mucopolysaccharidoses

Children with MPS should be managed by a multidisciplinary team with experience in the care of this patient group, and ideally in a hospital with pediatric intensive care facilities.²⁵ The risks of anesthesia must be carefully considered along with the anticipated benefits of the scheduled procedure and discussed with the child's family.²⁶ As demonstrated in our case series, patients with MPS who have received early treatment are less likely to present the same clinical challenges as those who remain untreated or received late treatment. They will have reduced risk and perioperative care requirements. The success of modern treatment options means that many children with MPS will now reach adulthood and present new challenges to a group of clinicians previously unfamiliar with this condition. This will present new challenges and risks, and continued guidance will be required from those with experience in the care of children with MPS.

A preoperative assessment should include evaluation of the child's cardiac and respiratory systems. Respiratory tract infections should be fully treated prior to anesthesia. Evidence of sleep apnoea should be sought including a review of sleep study results if available. The airway review should also include consideration of the cervical spine. The airway management plan must always anticipate a difficult airway, and an experienced anesthesiologist with appropriate equipment should be available.

Where appropriate, preoperative investigations may include full blood count and electrolytes, chest and cervical spine X-rays, electrocardiogram and echocardiography. Pulmonary function tests may be useful where there is risk of restrictive lung disease, sleep studies where a history of sleep apnoea is significant, and diagnostic imaging where cervical spine instability is a feature. Cervical spine instability most often occurs in MPS I (Hurler) & IV (Morquio) and these patients may benefit from assessment with flexion-extension images. Cervical canal stenosis may lead to compression in patients with MPS IV and should cervical myelopathy be suspected then it is best evaluated with magnetic resonance imaging.^{26–31}

It is useful to review previous anesthetic records, but progression of any airway and clinical findings is to be expected and the view at direct laryngoscopy may be significantly less favourable with increasing age. As we have found in our study, airway difficulty is significantly reduced in children with MPS I treated with

stem cell transplantation.^{16,17}

Sedative premedication should be used in these children judiciously. An anti-sialogogue may be useful to reduce secretions that may compromise airway management. Intravenous cannulation may be difficult due their coarse skin and tissue deposits.

The incidence of airway difficulties has been found to be 25%, with failure to intubate in 8% of children with mucopolysaccharidoses. In Hurler syndrome this is even higher, at 54% and 23% respectively.⁹ Our study demonstrated a 42% difficult airway rate with a 2% failed intubation rate in a tertiary referral centre staffed by pediatric anesthesiologists familiar MPS. Airway difficulties are due to a combination of factors including airway tissue deposition of glycosaminoglycans, a short, potentially unstable neck, poor joint mobility including the cervical spine and temporomandibular joints, macroglossia and micrognathia. Untreated, and sometimes despite treatment, all of these factors are expected to worsen with age.

Facemasks may not fit appropriately due to their abnormal facial features, and children with MPS often develop airway obstruction on induction. Obstruction may not be relieved by oropharyngeal airways, which may push the enlarged epiglottis downwards causing laryngeal obstruction. Nasopharyngeal airways are difficult to pass due to nasal tissue deposits. Some clinicians have advised forward traction of the tongue. Neck manipulation should be minimised, and where there is known atlanto-axial subluxation as occurs in Morquio syndrome (MPS IV), manual inline immobilisation should be employed.^{26, 29, 32}

The laryngeal mask airway has been found to be useful, and may be life-saving,³³⁻³⁴ however, tissue infiltration in the upper airway can make satisfactory placement difficult.³⁶ The endotracheal tube size required may be smaller than expected for age, and a range of sizes should be available.³⁷ The McCoy blade can be useful where an enlarged epiglottis is obstructing the view. Fibre-optic intubation may be useful where cervical spine instability exists or the view at laryngoscopy is poor.^{38, 39} As we have observed, video laryngoscopy is increasingly used in the airway management of MPS patients.⁴⁰ The equipment and staff able to perform emergency tracheostomy should be available.^{41, 42}

Post-obstructive pulmonary oedema has been described in patients with advanced disease. Pre-existing supraglottic obstruction may be compounded by an exacerbation of pre-existing glottic obstruction following tracheal extubation and this may contribute to the development of negative pressure pulmonary oedema. Of note pulmonary oedema can occur some hours after the obstructive event.⁴³ The timing of extubation should be carefully considered to ensure the patient is fully awake and dexamethasone given prior to extubation may reduce complications.

Postoperative care should take place in a monitored environment (high dependency or intensive care) and humidified oxygen and early physiotherapy are helpful. A multimodal approach to analgesia should occur. Regional anaesthesia may be challenging, but where practical, is a useful adjunct.^{44, 45} As the treatment options for this unique group of conditions evolves so will their medical needs. Similarly, as anaesthetic techniques, equipment and safety practices improve so will their perioperative requirements.

Conclusions

In summary, we present a case series of our experience with MPS patients over a 15-year period and review the current medical management of MPS and the challenges posed to anaesthetists caring for these patients. Our findings confirm previous reports of reduced airway difficulty in the MPS population treated with HSCT, although in our series, ERT did not make a difference in the incidence of difficult airway. Patients with MPS I who received HSCT were found to be less likely to have a difficult airway compared with those not treated with HSCT. We report an overall difficult airway rate of 42%, and observed that video laryngoscopy is increasingly used to secure the airway in this population. Despite known airway difficulty, a third of these patients underwent anesthesia for diagnostic imaging or minor procedures without airway instrumentation, and approximately half of all procedures were done on an ambulatory basis with the patients discharged home on the day of surgery.

Abbreviations

MPS: Mucopolysaccharidosis

HSCT: Hemopoietic stem cell transplant

ERT: Enzyme replacement therapy

DAW: Difficult airway

LMA: laryngeal mask airway

ETT: endotracheal tube

Declarations

Ethics approval and consent to participate: Ethics approval from the Research Ethics Board at the Hospital for Sick Children was obtained for the retrospective analysis of patient records and data. The requirement for individual consent for each patient was waived by the ethics committee.

Consent for publication: Not applicable.

Availability of data and materials: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

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Author contributions: GS conceived of the study, formed the study proposal and ethics application, collected and analysed data, and drafted the manuscript. CP prepared the study proposal and ethics application, collected data, drafted and reviewed the manuscript. GW prepared the study proposal and ethics application, collected and analysed data, and drafted and reviewed the manuscript. All authors read and approved the final manuscript.

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Tables

MPS Type	Name	Incidence	Inheritance	Defective enzyme	Coarse features	Short stature	Organo-megaly	Cardiac disease	Neurological deficit	Skeletal	Odontoid hypoplasia	Death (untreated)	Treatment options
1 H	Hurler	1:100,000	Autosomal recessive	α -L-iduronidase	xxx	xxx	xxx	xxx	xx	xxx	-	Child	HSCT +/- ERT
1 HS	Hurler/Scheie				xx	xx	xx	xx	x	xx	-	Early adult	ERT
1 S	Scheie				x	x	x	x	--	x	-	Normal	ERT
II Severe	Hunter	1:150,000	X - linked recessive	Iduronate 2-sulfatase	xxx	xxx	xxx	xxx	xxx	xxx	-	Child	ERT
II Mild	Hunter				xx	xx	xx	xx	--	xx	-	Late adult	ERT
III A - D	Sanfilippo	1:24,000	Autosomal recessive	Various	x	-	xxx	x	xxx	x	-	Teens to early adult	-
IV A & B	Morquio	1:100,000	Autosomal recessive	Galactosamine-6-sulfatase & β -galactosidase	xx	xxx	--	x	--	xxx	xxx	Adult	ERT
VI Severe	Maroteaux-Lamy	1:100,000	Autosomal recessive	N-acetylgalactosamine-4-sulfatase	xxx	xxx	xxx	xxx	--	xxx	xxx	Child	ERT
VI Mod	Maroteaux-Lamy				xx	xx	xx	xx	--	xx	xx	Teens to early adult	ERT
VI Mild	Maroteaux				x	x	x	x	--	x	x	Late adult	ERT
VII	Sly	1:250,000	Autosomal recessive	β -glucuronidase	xxx	xxx	xx	x	xxx	xxx	xx	Child to early adult	-

Table 1. Types and characteristics of mucopolysaccharidoses.

a	MPS I Hurler	MPS II Hunter	MPS III Sanfilippo	MPS IV Morquio	MPS VI Maroteaux-Lamy	Total
No. of patients (%)	29 (45%)	10 (15%)	13 (19%)	13 (19%)	1 (1%)	66
No. of anaesthetics (%)	188 (57%)	39 (12%)	33 (10%)	62 (19%)	10 (3%)	332
Ambulatory surgery	96	17	25	33	6	177 (53%)
Non-ambulatory surgery	78	19	7	27	4	135 (41%)
Emergency surgery	14	3	1	1	0	19 (6%)
Median age (yr)	4.5	6.9	9.6	6.1	2.5	5.5
Median weight (kg)	16	25	36	15	15	17.5
No. receiving HSCT*	19	0	0	0	0	19
Age (yr) of HSCT: median (range)	1.1 (0.3-1.9)	-	-	-	-	1.1
No. receiving ERT*	10	5	0	3	1	19
Age (yr) starting ERT: median (range)	3.5 (1-12)	11 (2-17)	-	13 (11-15)	2.5	5.6

Table 2. Patient demographics.

*3 patients received both ERT and HSCT

aSpecialty	% of total (345)
Radiology	45
- Diagnostic	28
- Interventional	17
Otorhinolaryngology	12
Dental surgery	9
Orthopedic surgery	8
Ophthalmology	8
General surgery	6
Spinal surgery	4
Neurosurgery	3
Cardiac surgery	1
Other (endoscopy, lumbar puncture, bone marrow aspirate, trans-esophageal echo)	5

Table 3. Procedure types.

	MPS I Hurler (n=29)	MPS II Hunter (n=10)	MPS III Sanfilippo (n=13)	MPS IV Morquio (n=13)	MPS VI Maroteaux- Lamy (n=1)	Total (n=66)
Difficult airways						
Total no. (% of MPS type)	10 (34)	9 (90)	2 (15)	7 (54)	1 (100)	28 (42)
No. of DAW treated with HSCT (n = 16) (%)	3 (19)	-	-	-	-	3 (19)
No. of DAW treated with ERT (MPS I=7, MPS II=5, MPS IV =3, MPS VI=1) (%)	5 (71)	4 (80)	-	3 (100)	1 (100)	13 (81)
No. of DAW treated with both (n = 3) (%)	0 (0)	-	-	-	-	0 (0)
No. of DAW in untreated (%) (Total untreated = 31)	2 (67)	5 (100)	2 (15)	5 (45)	-	14 (45)
Intubations						
						No. (% of all anesthetics or of all intubations)
All intubations (% of all anesthetics in MPS group)	67 (36)	21 (54)	18 (55)	26 (42)	6 (60)	138 (42)
Direct laryngoscopy	48	5	17	14	3	87 (63)
Assisted direct laryngoscopy (laryngeal pressure ± bougie ± two person-technique)	2	5	0	1	1	9 (7)
Fibreoptic bronchoscope	9	5	0	4	0	18 (13)
Video laryngoscope	7	5	1	6	2	21 (15)
Failed intubation	1	1	0	1	0	3 (2)
Other						
						No. (% of all anaesthetics)
Laryngeal mask airway (% of all anesthetics in MPS group)	60 (32)	9 (23)	6 (18)	11 (18)	0	86 (26)
Face mask only	25 (13)	8 (21)	5 (15)	4 (6)	1 (10)	43 (13)
Nasal prongs only	35 (19)	1 (3)	4 (12)	20 (32)	3 (30)	63 (19)
Regional only	1 (0.5)	0	0	0	0	1 (0.3)

Table 4. Airway management for all MPS patients.

Airway management technique		No.	%
ETT	Direct laryngoscopy	12	8
	Assisted direct laryngoscopy (laryngeal pressure ± bougie ± two person-technique)	19	13
	Fibreoptic bronchoscope	22	15
	Video laryngoscope	15	11
Laryngeal mask airway		32	23
Nasal prongs		22	15
Face mask		20	14

Table 5. Management of difficult airways. ETT = endotracheal tube