Limited maximal mouth opening in patients with spinal muscular atrophy complicates endotracheal intubation
An observational study

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Editor,

Hereditary proximal spinal muscular atrophy (SMA) is a severe autosomal recessive motor neurone disease caused by survival motor neurone (SMN) protein deficiency due to loss of function of the SMN1 gene. This causes the degeneration of spinal cord alpha-motor neurones leading to muscular weakness. The broad range of SMA severity is captured by the clinical classification system, distinguishing four phenotypes based on age at symptom onset and motor milestone achievement. SMA type 1 is characterised by infantile (<6 months) onset and the inability to sit, type 2 by onset between 6 and 18 months and the inability to walk, and type 3 by onset after 18 months. These patients may lose ambulation before adulthood. Type 4 represents adult-onset SMA.1

Weakness is most pronounced in axial, respiratory and proximal limb muscle groups. Susceptibility to respiratory infections due to a weak cough and need for scoliosis surgery before puberty are common complications in severely affected patients (i.e. SMA types 1 and 2).1 Brainstem motor nuclei are also affected, as reflected by limitations in maximal mouth opening (MMO) due to atrophy and fatty degeneration of the lateral pterygoid muscles, in addition to better known problems with chewing and swallowing. MMO limitations occur in 50 to 100% of patients depending on SMA severity, but patients are often unaware of this complication. Limitations in MMO progress with disease duration.2,3

Endotracheal intubation is a crucial and frequently used procedure in hospital care for SMA patients.1 Limitations in MMO have long been known to complicate this procedure,4,5 although previously it has not been studied specifically for SMA patients. We hypothesised that disease-specific MMO limitations in SMA may complicate intubation procedures as well. Therefore, we retrospectively reviewed all anaesthesiology reports of SMA patients currently participating in our national cohort study who received scoliosis surgery before the age of 18 years between 1991 and 2015 at our tertiary referral centre. We systematically extracted details of intubation procedures, including all comments on procedural complications. Patient characteristics, including SMA type and last documented MMO, defined as the distance between upper and lower front teeth plus overbite, were extracted from the national SMA registry.6 All participants had genetically confirmed SMA. Written informed consent for the use of these data was obtained. Ethical approval for our observational cohort study was obtained from the Medical Ethical Review Committee of the University Medical Centre Utrecht, Utrecht, The Netherlands (Chairperson E.M. van de Putte, MD, PhD) on 18 January 2010 (No. 09-307/NL29692.041.09). Details of all other methods and procedures were published previously.6

We identified 36 patients fulfilling inclusion criteria. Detailed anaesthesiology reports were available from 28 (77%) patients. Mean age at surgery was 8.1 years (median: 7.6 years, range: 3.9 to 15.5 years), mean disease duration 7.4 years (median: 6.7 years, range: 3.3 to 15 years). Five (18%) patients had SMA type 1c (i.e. the mildest variant of the SMA type 1 spectrum1) and 23 (82%) SMA type 2. Important complications of endotracheal intubation, explicitly attributed to MMO limitations, were documented in five (18%) cases (Table 1). These five patients had a mean MMO of 12 mm (median: 14 mm; range: 6 to 18 mm) at time of inclusion in our cohort study (Table 1). MMO data of 20 of the 23 (87%) patients without reported intubation problems was available and significantly larger ($U = 17$, $P = 0.027$, $r = 0.44$) at a mean of 26 mm (median: 26.5 mm; range: 8 to 45 mm). For further context, the range of MMO measurements in 174 patients with SMA participating in our population-based cohort study is summarised in Fig. 1.

Limitations in MMO are one of the symptoms in SMA caused by involvement of brainstem motor nuclei. Prevalence figures of reduced MMO, defined as 35 mm or less, have not been studied, but previous cohort studies have shown that MMO limitations may occur already in the 1st decade.5

We selected anaesthesiology reports during scoliosis surgery to investigate consequences of limited MMO in patients with SMA for medical care, as scoliosis is an important complication that occurs in all children with...
SMA types 1, 2 and the majority of 3a and requires surgery early in life.\textsuperscript{1} Our data suggest that these MMO limitations can severely complicate endotracheal intubation procedures in patients with SMA. This is important to recognise, as MMO limitations are a specific complication in SMA patients, the need for endotracheal intubation procedures is increasing and not limited to planned procedures. Patients with SMA types 1 and 2 are

\begin{table}
\centering
\caption{Complicated endotracheal intubation in spinal muscular atrophy patients with limited maximal mouth opening}
\begin{tabular}{|c|c|c|c|c|c|c|}
\hline
No. & SMA type & Year of birth & Age at surgery (years) & Year of surgery & Mouth opening in mm$^*$ & Complications during intubation and procedure-related comments$^b$ \\
\hline
1 & 1c & 1997 & 15.5 & 2012 & 15 & Limited mouth opening necessitated fibre-optic nasal intubation \\
2 & 1c & 1999 & 5.1 & 2004 & 8 & Nasal intubation was performed, due to very limited mouth opening \\
3 & 1c & 2002 & 5.3 & 2007 & 18 & Very difficult oral intubation due to limited mouth opening, eventually successful \\
4 & 2 & 1992 & 9.9 & 2002 & 6 & Fibre-optic oral intubation was performed, because of very limited mouth opening \\
5 & 2 & 2001 & 4.5 & 2005 & 14 & Difficult oral intubation due to limited mouth opening, even with a glidescope, causing temporary hypoventilation. Afterwards fibre-optic nasal intubation was performed \\
\hline
\end{tabular}
\end{table}

MMO, maximal mouth opening; SMA, spinal muscular atrophy. All intubation procedures were performed by paediatric anaesthesiologists and if fibre-optic nasal intubation was necessary, they were assisted by paediatric otorhinolaryngologists. a Measured in millimetres. \textsuperscript{b} As reported by the anaesthesiologist.

Fig. 1

(a) Maximal mouth opening measurements in millimetres in spinal muscular atrophy patients with complicated or uncomplicated endotracheal intubation prior to scoliosis surgery. For context, maximal mouth opening measurements in 174 spinal muscular atrophy patients in whom maximal mouth opening was measured as part of our national registry are shown in (b). Dotted horizontal lines represent a maximal mouth opening of 35 mm, the lower limit of a normal maximal mouth opening.\textsuperscript{2}

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frequently admitted to the ICU because of problems with clearance of secretions during airway tract infections. Under such circumstances, uncomplicated intubation procedures are vital for patient safety.

The available data suggest that the risk of a complicated intubation procedure particularly increases with an MMO of 20 mm or less (Fig. 1). It is important to note that this represents an estimate, as these values were not always obtained at intubation or pre-operative screening but often considerably later at enrolment in our registry. Considering that MMO progressively decreases over time, values above 20 mm may already be associated with complicated intubation procedures in SMA patients. We have previously observed that such severe limitations in MMO are not uncommon in children with SMA under 10 years of age.2

Recent introduction of the first SMA-specific therapy suggests that prognosis of SMA will improve in the coming decade. Limitations of MMO will, however, occur as long as SMA cannot be cured. Improved life expectancy of severe SMA may increase the need for medical treatments and procedures that require intubation.7 Awareness of this SMA-specific complication among paediatricians, intensive care specialists and anaesthesiologists is therefore important, as is the regular assessment of MMO limitations and documentation in patient files.

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Comparison of the ease of tracheal intubation using a McGrath Mac videolaryngoscope and a standard Macintosh laryngoscope in normal airways

A randomised trial

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Editor,

The McGrath Mac videolaryngoscope (Covidien France SAS, Paris, France) is a recent device with a blade similar to the Macintosh blade; it has no specific channel to guide the advancement of the tube. The McGrath Mac has been reported to be better than the Macintosh laryngoscope for successful intubation in difficult airways.3 The question arises of the place of the videolaryngoscope for routine intubation compared with the standard Macintosh laryngoscope and this was the aim of this trial.

The current monocentric randomised controlled trial was approved by the local Ethics Committee (No. 131060) and was registered on the ClinicalTrials.gov website (NCT02292901). Patients aged from 18 to 80 years undergoing elective surgery under general anaesthesia requiring standard endotracheal intubation were enrolled, whereas pregnant women, breastfeeding mothers, patients needing a rapid sequence induction, patients with ear–nose–throat surgery and with history of previous difficult intubation were excluded. Patients with potentially difficult intubation were also not included; this potential difficulty was defined as the presence of at least two of the following factors: diseases associated with difficulties in intubation or clinical symptoms of airway disease, snoring or obstructive sleep apnoea syndrome, short thick neck, limited mandibular protrusion, head and neck movement 80° or less, edentulous, thyromental distance less than 65 mm, interincisor gap less than 35 mm and Mallampati class more than II. At their admission in the operating room, patients were centrally

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