





TABLE III Complications in sitting position

| <i>Complications</i>            | <i>Patient age</i> |
|---------------------------------|--------------------|
| Bilateral brachial plexus palsy | 11 months          |
| Bilateral brachial plexus palsy | 11 years           |
| Macroglossia                    | 18 months          |
| C-1 Spinal cord infarction      | 5 years            |
| C-1 Spinal cord infarction      | 14 years           |
| Venous air embolism (VAE)       | 4 months           |
| VAE                             | 12 months          |
| VAE                             | 18 months          |
| VAE                             | 2 years            |
| VAE                             | 2 years            |
| VAE                             | 4 years            |
| VAE                             | 5 years            |
| VAE                             | 6 years            |
| VAE                             | 7 years            |
| VAE                             | 12 years           |

venous air emboli occurred in children 12 years of age or less.

In the prone patients, complications included an accidental extubation, one case with very severe venous bleeding, and one incident of venous air embolism (Table IV).

### Discussion

Our experience indicates that the most common problem seen clinically originates from the high incidence of spinal and foramen magnum stenosis. These patients present with sleep apnoea and nocturnal airway obstruction which is probably related to brain stem compression.<sup>6-8</sup> Patients with sleep apnoea tend to have apnoeic spells followed by periods of hyperventilation. Therefore, carbon dioxide retention has not been a problem. Most of the patients for suboccipital craniectomy were under 12 years old and pulmonary function was not evaluated. Other problems associated with spinal stenosis include chest deformity and upper cervical myelopathy.<sup>9,10</sup> Some of the patients have such severe spinal stenosis that they present with long tract signs and need decompression to preserve peripheral neurological function.

TABLE IV Complications in prone position

| <i>Complication</i>   | <i>Patient age</i> |
|-----------------------|--------------------|
| Accidental extubation | 18 months          |
| Venous air embolism   | 7 months           |
| Severe bleeding       | 9 years            |

Achondroplastic dwarfs classically have a large protruding forehead, a short maxilla, large mandible, and large tongue and fall into a group that would immediately alert the anaesthetist to a potential problem in airway management. Certainly, these facial features may lead to difficulties in obtaining a seal with a mask. In our experience, however, mask ventilation was not a problem and intubation has been without difficulty. There are two reports of difficult laryngoscopy and endotracheal intubation in the literature<sup>3,11</sup> and in both cases the difficulty was attributed to an inability to extend the neck. We have not seen this problem in our patients.

We were interested in the relationship between the appropriate endotracheal tube size when compared with patient's age and weight. While most formulae for establishing the appropriate-sized tube are based on age, it is our experience that weight is a far more valuable guide in these patients. Smaller diameter tubes were required in the majority of patients in our series than would have been predicted on an age basis calculation. This is in agreement with the findings of Walts<sup>3</sup> (Table II).

Because these patients have spinal stenosis it is very important to be able to examine them in the immediate postoperative period. We therefore selected an anaesthetic technique which would allow this to be accomplished. Patients who either had intravenous catheters in place, or were old enough to tolerate them being placed before induction, had intravenous inductions with thiopentone. Anaesthesia was maintained with muscle relaxants, and inhaled isoflurane and nitrous oxide in oxygen. In the younger patients inhalation inductions were conducted with halothane because of the ease with which children accept this technique. Once induction and intubation had been accomplished, isoflurane was substituted for halothane, because it theoretically allows faster recovery. None of the dwarfs showed abnormal response to non-depolarizing muscle relaxants, or other drugs.

These patients tend to have excess skin and subcutaneous tissue, which makes the establishment of intravenous lines more difficult. The placement of peripheral intravenous lines and arterial catheters did not present a significant problem. However, placement of central venous catheters for those in the sitting position proved more difficult. Dwarfs have short necks, and this, associated with



saliva and secretions over the adhesive tape which secured the tube. Since that incident we have secured our tubes using a waterproof adhesive plastic drape and supported endotracheal and ventilator tubing after turning the patient prone. One patient whilst prone also experienced venous air embolism. This position should offer protection from venous air embolism by eliminating the pressure gradient between wound and right atrium. However, as previously reported,<sup>18</sup> venous air embolism can occur whilst prone, with disastrous consequences.

In conclusion, we have presented our experience in anaesthetizing 27 achondroplastic dwarfs for 36 procedures. We found no difficulty in airway management during induction, and found that weight was the best guide for assessing the size of the endotracheal tube. We experienced some difficulty in positioning the very small children in the sitting position as no commercial chair is available. We further believe that the brachial plexus palsies, macroglossia and brain stem infarction are associated with this positioning problem, and we have recommended how to best avoid these complications in the future. We have pointed out the advantages and disadvantages of the sitting and the prone position and recommend that because of our experience, extreme care be taken to avoid venous air embolism if the procedure is undertaken in the sitting position.

## References

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## Résumé

*Au cours de trois dernières années, 36 anesthésies ont été administrées à 27 patients atteint nanisme achondroplasique. Vingt quatre patients ont subi une crâniectomie pour sténose du foramen occipital. Seize de ces opérations ont été accomplies dans une position assise avec neuf incidents d'embolies gazeuse, tous survenant chez des patients âgés de moins de 12 ans.*

*Six complications majeures sont survenues: deux infarctismes de la moelle épinière au niveau de C-1, deux plexus brachial, une macroglossie sévère, et une extubation accidentelle.*

*L'accès intraveineux du patient avec un nanisme achondroplasique est difficile à cause de l'excès de peau et de tissus sous-cutané.*

*La conduite de maintien des voies aériennes et la laryngoscopie n'étaient pas difficile et on a trouvé que la grosseur du tube endotrachéal est mieux prédite par le poids du patient plutôt que son âge.*

*Les pertes sanguines ont été de  $38 \pm 9 \text{ ml}\cdot\text{kg}^{-1}$  en position couchée ( $n = 8$ ) et  $18 \pm 4 \text{ ml}\cdot\text{kg}^{-1}$  en position assise ( $n = 16$ ), et était en relation avec la position chirurgicale plutôt qu'au nanisme.*

*Nos données indiquent que les complications surviennent plus fréquemment en position assise celles ci sont dangereuses et toutes les précautions doivent être prises afin d'éviter leur survenue.*