ANESTHETIC CONSIDERATIONS IN DOWN SYNDROME

Down Syndrome, or trisomy 21, is the most common chromosomal anomaly, occurring in 1/700 to 1/1100 of live births. The children are disabled by mental retardation (100% have IQ's below 65), seizure disorder (2-9%), congenital heart disease (50%), cervical spinal laxity, and an increased incidence of neonatal GI lesions, leukemia, and respiratory infections.

The cardiac lesions are divided between VSD (30-40%), endocardial cushion defects (AV canal) (32%), and other defects associated with pulmonary hypertension. The combination of intracardiac shunts and pulmonary infections leads to the early development of cyanosis and Eisenmenger's syndrome. Open heart surgery for these children was often not performed prior to the last decade, but now most lesions are corrected in infancy. A thorough preoperative cardiac evaluation is necessary in all patients. Meticulous air bubble precautions and SBE prophylaxis is also warranted (see section on anesthesia for congenital heart disease).

As mentioned above, immunodeficiency and recurrent pulmonary infections combined with heart disease and craniofacial hypoplasia increases the risk of difficult oxygenation during the induction of anesthesia. These children may have a high arched palate, small upper face and nose, as well as a large tongue. To some degree, the joint laxity balances these factors so that intubation is usually not difficult. Care should be taken not to hyperextend the neck, as cervical spinal dislocations have occurred. Ten to thirty percent of all children with Down Syndrome will have cervical spinal stenosis or atlanto-axial subluxation on C-spine films, although only 1-2% will become symptomatic. These symptoms include neck pain, torticollis, quadraplegia, clonus or weakness in the lower extremities. In the past, we have required all patients with Down syndrome to have a preoperative C-spine survey (by age 2-3 years) , due to reports of symptomatic atlanto-axial subluxation after anesthesia - perhaps due to hyperextension of the neck during intubation, or perhaps related to generalized muscle relaxation. The issue was always.. what do you do with abnormal films in the face of no symptoms.. and what about normal films.. are these patients not at some risk, as well?? At the present time, we do not *require* these films (although CDC patients usually have them on file) and treat all Down Sx patients with positioning care.

Approximately 25% of children require an ETT that is 1-2 sizes smaller than expected, at times (but not always) due to generalized smaller size. Adults may also have relatively smaller tracheas. Postoperative stridor and other respiratory complications are more common than in the general population. Postoperative airway obstruction may also be due to macroglossia and poor pharyngeal tone.

In addition to the problems cited above, gastrointestinal defects are common. Imperforate anus, tracheoesophageal fistula, and duodenal atresia are the most common.

CNS abnormalities in patients with Down syndrome may be associated with altered concentrations of neurotransmitters, such as serotonin, dopamine, norepinephrine and acetylcholine. Therefore, the anesthesiologist should be very careful in the administration of sedatives and narcotics to these patients.

There have been reports of exaggerated response to atropine, with tachycardia and cycloplegia.

In summary, most anesthesiologists will encounter these patients in their practice, as life expectancy and surgical correction of the multiple lesions are becoming more common. Thus, even the non-pediatric anesthesiologist should have a thorough knowledge of the problems associated with Down syndrome.

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