

The FLK for tonsillectomy

Beeton A

Private Practice Johannesburg

With affiliation to University of the Witwatersrand, Anaesthetic Department

Keywords:

What is a FLK?

A child with morphological features at variance from societal perceptions of normality (too big, too small, too fat, too floppy, too stiff, too hairy, too distorted, funny face). The more "abnormal" features present, the more likely that the child has a syndrome. However, FLKs do not have to be syndromic. They may instead have malformations, disruptions, deformations, dysplasias, associations, complexes or sequences – all of which may be relevant to anaesthesia. The aetiology may be genetic or acquired. 3-7% of children are born with one or more congenital abnormality. 75% of these affect the cranio-facial area. Frequently, parents arrive at the pre-operative interview with literature related to the child's condition. Ignore this at your peril!

What features may point to dysmorphism?

- Growth abnormalities / height / weight / asymmetry
- Milestones
- Head shape and size / fontanelles / ridges
- Hair – colour / texture / HAIRLINE
- Eyes – colour / shape / SLANT / SPACING
- Ears – shape / POSITION / ROTATION
- Nose – shape / size / clefts
- Midface – flat / hypoplastic
- Mouth – size / shape / palate / teeth
- Chin
- Tongue – size / mobility
- Neck – length / shape / mobility
- Chest – symmetry / shape / nipples / spacing
- Heart – rate / rhythm / murmurs
- Abdomen – shape / visceromegaly / hernias
- Genito-urinary – development / ambiguity
- Neurological – mental state / tone / muscle strength, size & definition / reflexes / gait
- Back – deformities / sacral dimples
- Peripheries – symmetry; hands & fingers (creases, shape & number of fingers); club and flat foot; limb length / proportion abnormalities
- Skin – naevi / cafe au lait spots / rashes



Figure 1: Common facial measurements

(1) Interpupillary distance, (2) inner canthal distance, (3) outer canthal distance, (4) interalar distance, (5) philtral length, (6) upper lip thickness, (7) lower lip thickness, and (8) intercommisural distance

Patterns of dysmorphism

- Children of funny looking parents – does not exclude an abnormality / syndrome
- Recognisable syndromes with known associations e.g. Down's, Pierre Robin
- Definitely syndromic but unclear as to diagnosis – ideally get a diagnosis and look it up but if unsure, check heart and airway; consider TIVA; avoid suxamethonium and NDMRs; assume opioid sensitivity; supported ventilation; longer post-op observation
- Branchial arch abnormalities – ears and airways
- Big, flabby, slow kids with mental retardation
- Myopathic kids – tone problems; featureless faces
- Obvious mucopolysaccharidoses
- Adenoidal facies. The commonest and most underestimated source of peri-operative difficulties in tonsillectomy patients.

Importance of the dysmorphic child to the anaesthetist

- Awareness and relevance of associated abnormalities – esp heart, obstructive sleep apnoea and pulmonary hypertension
- The difficult airway

- MH associations
- Drug sensitivity
- Difficult vascular access
- Psychological issues

Associated conditions

Cardiac disease

Valvular disease, structural defects, cardiomyopathy, conduction defects, hypertension, cardiac failure and pulmonary hypertension may all be features of dysmorphic children. Not all dysmorphic children are, however, at risk for cardiac disease.

Who requires further investigation?

- Children whose history and examination suggests possible cardiac disease – effort tolerance, cyanosis, murmurs, arrhythmias, cardiac failure, poor feeding, frequent respiratory infections, poor growth
- Primary myopathies (dystrophies, mitochondrial disorders and storage diseases)
- Where the syndrome is frequently associated with cardiac disease e.g. Down's

Sleep apnoea and pulmonary hypertension

- Adenotonsillar hyperplasia is the commonest cause of sleep disordered breathing
- Other important causes include many of the cranio-facial dysmorphic syndromes characterised by midface hypoplasia, micrognathia and macroglossia (Down's; Pierre Robin; Treacher Collins; Apert; Crouzon) or obesity (Prader Willi; Beckwith Wiedemann; Lawrence Moon Biedl)
- Complications include sleep and behavioural disturbance, airway obstruction related to anaesthesia and sedation, hypoxaemia, RV hypertrophy and failure and dynamic (later fixed) pulmonary hypertension
- Factors with highest risk for complications
 - Age < 3
 - Severe OSA
 - Failure to thrive
 - Cardiac complications
 - Obesity
 - Prematurity
 - Recent URTI
 - Cranio-facial abnormalities
- General anaesthetic principles
 - Defer patients with acute URTIs
 - Avoid sedative premedication. Dexmedetomidine acceptable.
 - Gas induction with CPAP +/- naso-pharyngeal airway
 - Intubation and ventilation with small dose of NDMR
 - Awake extubation with indwelling naso-pharyngeal airway in severe cases
 - Balanced analgesia (opioid light) – LA, paracetamol, steroid, NSAID
 - Prolonged monitoring – high care for high risk cases

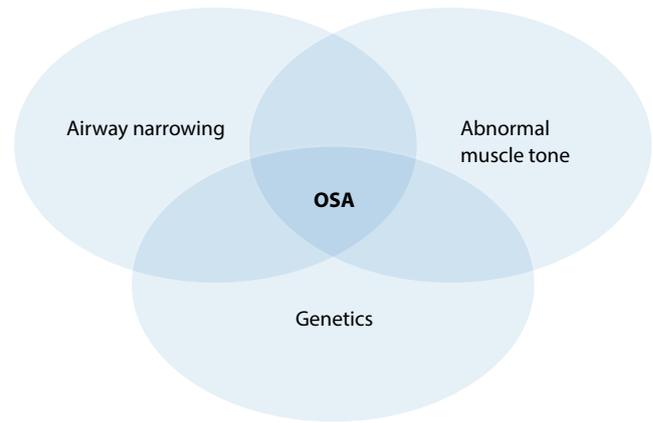


Figure 2: Pathophysiology of paediatric obstructive sleep apnea syndrome. The difficult influences on the airway are additive. Adapted from Marcus, *Respir Physiol* 2000;119:143-54

Other

- Renal disease e.g. polycystic kidney disease
- Skeletal abnormalities making positioning difficult or predisposing to restrictive lung disease
- Respiratory problems – restrictive lung disease; chronic reflux; recurrent lung infections; inherent disease e.g. cystic fibrosis
- Metabolic abnormalities – may be primary e.g. PKU or secondary e.g. hypoglycaemia in Beckwith – Wiedemann syndrome. Drug pharmacokinetics may be impaired
- Neuromuscular – patients with myopathies may be sensitive to neuromuscular blockers or develop sudden cardiac arrest

Malignant hyperthermia

There are relatively few myopathies unequivocally and predictably associated with the development of MH in response to suxamethonium or volatiles (King Denborough syndrome; central core disease and Evans myopathy). These children tend to be floppy, have muscle wasting, skeletal abnormalities, ptosis and strabismus. Numerous other myopathies e.g. Duchenne, storage diseases and mitochondrial disorders have, in the past, been associated with MH but, mechanistically, their reactions to volatiles and suxamethonium differ from MH and have been reclassified as AIR (anaesthesia induced rhabdomyolysis). The attack of hyperkalaemia, gradual hyperthermia and rhabdomyolysis settles on withdrawal of the volatile agent, but suxamethonium remains contra-indicated. Danrolene is generally not required. When in doubt, floppy or myopathic children should receive TIVA or TCI with propofol. One word of caution – children with mitochondrial myopathies are at increased risk for the propofol infusion syndrome – at lower doses and shorter infusion periods than is the rule. There is no real association with MH in this group, so, for prolonged procedures, a volatile technique is acceptable.

The difficult airway

Certain conditions are always associated with difficult airway access – Pierre Robin sequence; Treacher Collins syndrome; Goldenhar syndrome. These all present obvious features of difficult intubation including micrognathia, macroglossia, clefts,

high arched palates, abnormal dentition and a high, anterior larynx. Other dysmorphic children – particularly those with low slung and misshapen ears may present with unexpected airway problems – related to abnormalities of neck and jaw movement, mouth and tongue size and shape, choanal atresia, dentition or other airway pathology. A clue may be a history of snoring and recurrent URTIs – hence presentation for tonsillectomy.

A difficult intubation should be suspected in all dysmorphic children. A good pre-operative airway assessment may still miss choanal, laryngeal and epiglottic abnormalities as well as cranio-spinal instability (e.g. in Down's, Morquio's and Hurler's syndromes) or immobility (e.g. Klippel Feil), that may impact intubation. A properly stocked difficult intubation trolley should be present in theatre. Appropriate sizes of airways, LMAs and ETT tubes should be available. A video-laryngoscope with a paediatric blade is invaluable. In general, a spontaneously breathing volatile induction is recommended to allow laryngoscopy before administration of relaxants. Where volatiles are clearly contra-indicated, a variety of intravenous techniques have been described to facilitate laryngoscopy whilst retaining spontaneous respiration – dexmedetomidine alone or a combination with ketamine (with glycopyrrolate) – offering the best chance of avoiding apnoea.

Drug sensitivity

As a general rule, syndromic children are more than usually sensitive to opioids, including weak oral opioids, and neuromuscular blocking agents. Responses to benzodiazepines may often be paradoxical.

Difficult venous access

Several conditions are associated with difficult vascular access due to limb deformities, obesity and thickened subcutaneous layers. This may lead to prolonged anaesthesia and hypothermia due to repeated attempts to cannulate in an uncovered infant. Attention maintaining temperature is crucial.

Psychological issues

Dysmorphic children (and their parents) are often very agitated about hospitalisation, anaesthesia and surgery. They have frequently had previous traumatic experiences as they, particularly those with cranio-facial abnormalities, are regular visitors to theatre. Many dysmorphic syndromes are associated with mental retardation, attention deficit hyperactivity disorders or autism - all of which may produce a hysterically uncooperative patient.

The options are attempting to establish rapport with the child, presence of a calm cooperative parent, premedication, IM ketamine (+/- midazolam) or a veterinary approach – forced volatile anaesthesia with physical restraint. The last mentioned approach is virtually impossible in bigger mentally impaired children and guarantees an emergence reaction in 100% of cases. Midazolam 0.5 mg/kg 15 – 30 min pre-op is the most frequently used premedication, but is unpredictable and may produce dysphoria. Oral or nasal dexmedetomidine (1 – 4 µg/kg, 30 – 60 min pre-op) would appear to be the most predictable current option.

Extensive reference was made to "Anaesthesia for the Dysmorphic Child", the MMed thesis of Dr JL Taylor, UKZN: 2009. Further references available on request.