Dentistry + paediatrics + Maxillofacial Surgery = not so ordinary

I know there are many of your readers who have taken the opportunity to work at least at some point in their careers in a hospital, but not everyone. Even fewer have had the chance to work at one of only three Paediatric Craniofacial units in the UK. I write to you hopefully to give the readership an insight into the Paediatric Maxillofacial world. Working in such a unit could be described as working with children with complex medical needs, complicated medical histories, complex treatment plans, at times with difficult behavioural issues, with acute and urgent needs. But I would simply describe it as the most rewarding job I have done. Why? Well, as a dentally qualified professional in a medical environment it is a challenge. There is a quick and often brutal learning curve both clinically and academically. Working with children brings in another challenging factor; they can be both predictable and unpredictable and the clinical decisions can be obvious or completely unknown territory.

Imagine being 12 years old and having to take a decision to have your permanent teeth removed because they've become too painful and mobile to eat. What will your friends say at school? Actually, will you even be able to go back to school? A real patient with Papillon-Lefèvre Syndrome.

What about being a 14-year-old girl having to undergo a mandible resection because of a large tumour. Not your fault, but what other choice do you have? It's already disfiguring but what will the outcome be? A real case presenting with a psammomatoid ossifying fibroma.

What about a 5-year-old with a fatal heart condition who desperately needs some teeth taking out so that he can go on to have his cardiac operation. Where would you start with managing his medical needs? How could you explain to his parents the real risks of their child not surviving a general anaesthetic? A real patient on a cardiac transplant list with a failing Fontan Circulation.

The knowledge and experience gained in this paediatric environment has profoundly changed my practising career. The 'not so ordinary' will stay with me as I go further in speciality training.

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Keep an eye out for the bifid uvula: yes please – as it is not always just an isolated congenital anomaly

Bifid uvula is typically associated with submucous cleft palate but has often otherwise been considered innocuous, apart from dental anomalies as in your recent paper.[1] The generally accepted definition of a submucous cleft palate is the triad of bifid uvula, notching of the hard palate, and muscular diastasis of the soft palate, but it can occur in the absence of a bifid uvula; most patients with bifid uvula, however, have a submucous cleft.[2] Bifid uvula is more common in males than in females. Rare associations include the occurrence of bifid uvula in Loeys–Dietz syndrome[3] and inflammatory linear verrucous epidermal nevus syndrome.[4]

It is therefore important also to exclude a diagnosis of Loeys–Dietz syndrome, a recently recognized autosomal dominant condition associated with increased transforming growth factor (TGF beta) signalling in blood vessel walls, predisposing to thoracic aortic aneurysm (TAA) dissection.[5] Loeys–Dietz syndrome presents with some features similar to Ehlers-Danlos/Marfan syndromes such as:

- Bifid uvula and/or submucous cleft palate;
- Arterial aneurysms;
- Bicuspid aortic valve;
- Osseous anomalies (eg scoliosis , club foot);
- Hypertelorism;
- Allergies.

Complications of Loeys–Dietz syndrome include a predisposition to arterial rupture, cervical spine instability and, for women, increased risks during or after pregnancy from aortic dissection or uterine rupture.

Thus a thorough clinical examination and accurate medical history, and awareness of the systemic implications of apparently innocuous conditions, such as bifid uvula, allows for a possible diagnosis of a syndrome which can have serious implications for systemic health. Investigations, including echocardiography, head to pelvis 3D CT angiogram scan for evidence of vascular pathology, and mutation testing for Loeys–Dietz syndrome, may be indicated and the angiotensin II type 1 receptor antagonist, losartan, might be considered for prophylaxis.

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Placement of Class II composites

I refer to Alix Furness's letter on the placement of Class II composites without the use of a matrix band (December *Dent Update* 2014; **41**: 933). I was attracted to the article because of its heading but, as I continued to read it, I realized that the technique alluded to was unconventional to say the least.

I was intrigued since I, too, have been qualified for well over 30 years and have been to many hands-on courses for placing posterior composites, including some presented by Professor Burke. I have spent a professional lifetime trying to perfect my technique and have never attempted doing them free-hand, much less had it recommended by anyone as an evidence-based procedure.

I presume that there is a reason for this.

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