Access to special care dentistry, part 8. Special care dentistry services: seamless care for people in their middle years – part 2

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VERIFIABLE CPD PAPER

This article about special care dentistry in the middle years considers people who have Down's syndrome and cerebral palsy and those who have cardiac and respiratory disease. The increased life expectancy of people with Down's syndrome, currently 50-60 years, is reflected in the changing population profile and needs of these individuals. The preventive and dental treatment of most people with Down's syndrome and cerebral palsy can be met in general dental practice. However, those people with profound disability, anxiety or learning disability may require either a shared approach to care or referral for specialist care. Cardiac and respiratory disease occur commonly in the general population both in middle and older age groups and the dental team will meet increasing numbers of people with these conditions. The procedures and drugs used in dentistry can aggravate heart disease and it is important that the dental team are aware of the common cardiac conditions and their management, as well as how to best manage the oral care of this group. Also, they have a role to play in the provision of oral health advice, smoking cessation and dietary advice. This is particularly important as poor oral hygiene has been linked to respiratory pathogen colonisation and dental plaque may act as a reservoir for aspiration pneumonia in susceptible individuals.

IN BRIEF

- Most dental treatment for adults with Down's syndrome should be possible in primary dental care.
- Higher levels of oral disease can occur among people with cerebral palsy.
- It is important that the dental team are aware of the common cardiac conditions and their management.
- The dental team has a role to play in the provision of oral health advice for patients with respiratory disease.

PRACTICE

ACCESS TO SPECIAL CARE DENTISTRY

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DOI: 10.1038/sj.bdj.2008.850
©British Dental Journal 2008; 205: 359–371

This second article on seamless care for people in their middle years considers two conditions which have traditionally been considered with childhood and young adult conditions, and two conditions that have traditionally been associated with older people. The first two – cerebral palsy and Down's syndrome – are included in this article to reflect the increasing life expectancy of people with these conditions and the subsequent change in their population profile and needs. The latter two conditions – cardiac and respiratory disease – now occur commonly in middle age as well as in older age and the dental team will see increasing numbers of people with these conditions.

1. CEREBRAL PALSY

Cerebral palsy (CP) is an umbrella term encompassing a group of non-progressive neurological and physical disabilities caused by damage or a lesion to a child's brain early in the course of development, either *in utero*, during birth or in the first few months of infancy.[1] The damage to the brain is caused mainly by hypoxia, trauma and infection but genetic and biochemical factors have also been suggested.[1] Pre-natal risk factors include pre-eclampsia, irradiation, a maternal age of less than 20 or over 35, and infections such as cytomegalovirus, rubella and syphilis. Peri-natal risk factors include trauma, breach birth or prolonged delivery.[1,2] Damage may also be caused post-natally following infections such as encephalitis and meningitis during infancy. Other risk factors include cerebral ischaemia, haemorrhage and hypoxia secondary to trauma, respiratory distress, hypothermia or hypoglycaemia.[1,2]

Cerebral palsy is the most common congenital cause of physical impairment,[1] with an incidence of approximately 2-2.5 per 1,000 live births in developed countries.[2] Primarily it is a disorder of voluntary moment, which results in a wide spectrum of disability ranging from virtually unnoticeable physical impairment. It may affect only one limb (monoplegia), both lower limbs (paraplegia), one upper and one lower limb on the same side (hemiplegia) or all four limbs equally (quadriplegia).[1] There are four main types of CP (Table 1), the

features of which are governed by the area of brain damage.[2,3]

Diagnosis

Diagnosis is usually made from clinical signs, such as weakness in one or more limbs, abnormal gait with one foot or leg dragging, excessive drooling or difficulties in swallowing and poor control over hand and arm movement. Other impairments which may accompany CP include visual, hearing and speech impairments, epilepsy, drooling and learning disability.[4] Less than 50% of individuals with CP have a learning disability and indeed many people are highly intelligent and well educated, though severely impaired speech due to dyspraxia or dysphasia and sensory impairments can mislead some unwary observers.[1] Although CP is a non-progressive disorder, other secondary complications may occur and can include respiratory complications, secondary digestive system problems (reflux and constipation), bladder infections and kidney infections, skin problems on pressure areas and peri-orally from drooling, and musculo-skeletal problems such as arthritis, dislocations and deformities.[5]

Table 1 The features of the four types of cerebral palsy (CP)

Type of CP	Percentage of all CP types	Site of cerebral damage	Features
Spastic	55%	Cortex	Increased muscle tone and contractions of affected limbs Complete/partial loss of control of muscle movement Difficulties with head support Difficulties with control and balance Falls easily
Athetoid/ Dyskinetic	20-25%	Basal ganglia	Smooth writhing movements as muscle tone fluctuates from increased to decreased tone Constantly in motion High energy requirements Movements exaggerated if anxious or an effort is made to control them
Ataxic	10%	Cerebellum	Disordered short, jerky movements Difficulties with balance Difficulties with walking and sitting straight Requires time to execute changes in position Compromised co-ordination
Mixed	10-15%	Some or all of the above	Some or all of the above Tremor of all or part of the body Hypotonia of muscles Inability to stand Inability to raise the head

Treatment

While there is no cure, therapy can help children, adults and their families manage the problems that cerebral palsy presents. 'Bobath therapy' is very popular and is a trans-disciplinary approach using specialised handling and posture techniques to encourage more controlled patterns of movement. Combined with physiotherapy, occupational therapy and speech and language therapy it can change the clinical presentation of CP.[5] Splints, orthopaedic surgery and medications such as muscle relaxants are used to relieve muscle stiffness and to reduce pain and contortions.[1] Between 25-30% of people with CP have epilepsy and take related drug therapy.[1] Dietary advice is required where nutrition or swallowing is compromised.[4] Life expectancy in CP has

Fig. 1 Wheelchair designed to provide maximum support and independence for a woman with cerebral palsy

Fig. 2 The personal assistive devices near to 'hand' for a woman with cerebral palsy

increased significantly in the last decades, however respiratory infections are common and aspiration pneumonia is a major cause of death.[1] Wheelchair design and assistive devices can help to provide a degree of independence (Figs 1 and 2).

Oral and dental features

People with CP will encounter the same oral and dental disease as the rest of the population, however there are additional factors such as access to dental care and support in carrying out daily living activities, which can result in higher levels of untreated disease and tooth loss.[4] Scope, the national voluntary and political body for CP, works actively on campaigns to 'get equal' and make 'rights a reality'. At the time of writing it was running an online campaign seeking out 'disablism', which it describes as 'discriminatory, oppressive or abusive behaviour arising from the belief that disabled persons are inferior to others'.[2] It will only be a matter of time before their active and ongoing research highlights the inequality in oral health and acts accordingly.

There are many potential causes of increased risk of dental disease in CP. They include:

Developmental abnormalities – the maxillary arch is frequently tapered or ovoid and the upper incisors may be labially inclined, making oral hygiene difficult.[1] The incidence of malocclusion is high and delayed eruption, poor oromuscular co-ordination, lack of adequate lip seal and oral habits of tongue thrust contribute to this.[1,3,4]

Uncontrolled movement – characteristic symptoms of the movement disorder may be observed in the orofacial and cervical muscles,[6] including spasticity of the temporomandibular joint (TMJ) musculature.[3] Facial grimacing, dysphagia and swallowing difficulties are common[3] and jaw dislocation due to spontaneous subluxation may occur.[1,4]

Bruxism and tooth wear – these are common in CP, especially in those individuals with athetoid CP.[3] Loss of tooth tissue may be exacerbated by erosion due to gastro-oesophageal reflux, which is also common.[1,4]

Periodontal disease – is reported in a high proportion of people with CP which affects their upper limbs and manual dexterity. Pre-disposing factors to periodontal disease in this group include mouth breathing, gingival hyperplasia secondary to the use of phenytoin for the treatment of epilepsy[7] and increased food retention which is exacerbated by difficulties in oral self-care and plaque removal.[3,4] The increasing use of 'peg' (percutaneous endoscopic gastronomy) feeding has helped improve the nutritional status of patients with swallowing difficulties, but the need for regular and meticulous oral hygiene has not been addressed[4] even though Dicks *et al.*[8] have shown that calculus formation is significantly more rapid in tube-fed patients.[8] This is important as poor oral health in patients with dysphagia has frequently been associated with the development of aspiration pneumonia.[9] There is good evidence that improved oral hygiene and frequent professional oral healthcare reduces the occurrence or progression of respiratory diseases.[10]

Caries – the risk of caries is increased by a number of factors related to CP and its treatment, for example reduced chewing and swallowing ability, the tendency for food to be retained in the mouth,[4] malocclusion and mechanical and physical difficulties in removing plaque. In the absence of effective oral hygiene procedures, individuals with feeding difficulties who use dietary supplements and laxatives with a high sugar content can develop extensive levels of caries rapidly.[4]

Fractured teeth – the increased likelihood of falls[3] and seizures[1] amongst people with CP means that fractured teeth are more likely than in the general population.

Xerostomia – dry mouth secondary to the use of medication to control seizures is undoubtedly a causative factor of oral disease.[7] However, studies have shown that even when not taking such medication, people with CP have a lower than normal salivary flow rate, lower pH and reduced buffering capacity, further increasing their risk of oral disease.[11] Exacerbated by mouth breathing, crusting mucous deposits are commonly seen on the palate and soft tissues.[4]

Drooling – problem drooling affects up to 58% of children with cerebral palsy[12,13] and although the incidence is lower in adults, for many individuals it is severe enough to interfere with daily social and practical functions.[13,14] It is not caused by hyper-salivation, but is due to impaired swallow and poor control of the orofacial musculature[15,16] and can be exacerbated by malocclusion, postural problems, dental caries and an inability to recognise salivary spill.[17]

Fig. 3 A woman with cerebral palsy using a mouth-held device to aid independence

Lip trauma – is a condition associated with individuals who have a profound neurodisability[18,19] and has been reported in people with cerebral palsy.[20,21] The bite reflex occurs pathologically in this group and is often a result of facial hypersensitivity, anxiety and poor head position.[19,22]

Seamless care

The management of oral health needs to be embedded into the general care plan of every individual with CP. While at times the dental team may be on the periphery of the multidisciplinary team, at others they are integral and can play a significant role in improving the quality of life for people with CP. Some of the ways they contribute to this include:

The maintenance of independence. If an individual relies on mouth-held devices to carry out certain activities or to assist communication, maintaining good oral health is critical to retaining independence (Fig. 3).

The management of xerostomia and erosion. Strategies for management of xerostomia will be discussed in more detail in the next paper in the series. Characteristic problems such as mucus or crusting deposits on the tongue, in the palate or on the teeth may be prevented by the use aqueous lubricating gel both peri- and intraorally. Deposits may be removed by gentle brushing with a soft toothbrush or swab dipped in mucolytic solvent, for example Bisolvon 2 ml/mg (bromhexin).[23] Erosion and hypersensitivity may be addressed with topical fluoride application as outlined in article 4,[24] with the use of gel, varnishes or mousse preparations being more appropriate than mouthwash in those patients with dysphagia.[4,23]

Management of bruxism. Bruxism reflects a multi-factorial interaction of anatomical, physiological and psychological factors and, not surprisingly, is more common in patients with movement disorders and learning disability.[1] Goals of treatment are reduction of clenching, reduction of any associated pain and prevention of further tooth damage. Traditional treatment options including use of occlusal adjustment maybe of limited success,[1] and first-line management for people with CP is to ensure that trigger factors (such as caries and sharp teeth, pain, poor posture and stress) are reduced.[22] Appliance therapy, even if the individual can tolerate it, may be compromised by the difficulty in obtaining impressions and the risk of compromised swallowing and airway protection. Rather than using alginate, Milwood *et al.*[19] advocate the use of silicone putty impression material (with removal from the mouth before final set to avoid locking into undercuts and interdental spaces) and props to aid opening[19] during impression taking. Other reported successful management strategies include relaxation of facial and TMJ muscles by massage techniques,[4] cryotherapy[25] and the use of medications such as gabapentin[1] and botulinum toxin[26] which are best managed by a specialist multidisciplinary team.

Management of drooling. Drooling management is a complex clinical problem that involves a multidisciplinary team approach.[16] Strategies include oromotor therapy, appliance therapy and behaviour modification in childhood;[14] or radiotherapy, drug therapy and surgery for adults.[16] Treatment should progress from the least invasive modality to the most invasive. Behavioural modifications coupled with oromotor therapy seems to be the treatment of choice for children with CP.[27] However, where drooling persists into adulthood, it can have a significant impact on quality of life[17] as it impairs masticatory function and speech and increases the likelihood of maceration and peri-oral infections.[15,17] Furthermore, drooling can be embarrassing and detrimental to peer bonding.[15,17] Overall it seems that surgical and pharmacological approaches are entirely empirical and there is no evidence to suggest a more successful outcome for any particular approach.[14] Reversible options include scopolamine patches[1] and botulinum toxin injections into the salivary glands.[26] The disadvantage of botulinum toxin is its short duration of action (two to six months) which means repeated injections are required. This is a dilemma if patients who are unable to co-operate need repeated general anaesthesia (GA) for this purpose. Also, the highest safe

Fig. 4 Transfer from wheelchair to dental chair by hoist for a patient with cerebral palsy

Fig. 5 Man using a mouth-held device to operate his computer, which is used for communication

dosage and the long-term complications are still unknown.[15,16,28] Surgery, although non-reversible, has a high success rate[15,16] with preference given to more conservative procedures, such as sub-mandibular duct relocation.[15,16]

Treatment of drooling, whether pharmalogical or surgical, results in reduced salivary flow and the patient will always be more susceptible to dental caries.[16] Regular dental examination and extra preventive measures, as detailed in article 4 of this series, are therefore mandatory for patients undergoing this type of therapy.[24]

Management of lip trauma. Various oral appliances have been used in an effort to prevent trauma and promote healing of lesions.[18,20,21,29,30] The drastic solutions of extracting teeth and orthognathic surgery to create an anterior open bite have been used in isolated cases.[31] The removal of teeth does not usually solve the problem as trauma is relocated to a different site once the lip support is reduced following extractions.[19] A dentist-led multidisciplinary team acknowledged that current clinical management options are limited, often ineffective, and require further development.[22] Numerous factors affect the choice of treatment, with patient co-operation, access to the oral cavity and wishes of the individual and family being the most limiting factors.[22]

Systemic desensitisation through touch, using techniques common amongst speech and language therapists, has been shown to be useful in the prevention of bites and to facilitate impression-taking for fabrication of hard acrylic bite guards, lip bumpers or soft vacuum moulds.[22] Opening of the mouth may require assistance or use of one of the aids or props outlined in article 1 of this series.[32] Similarly, the use of botulinum toxin[26] and muscle relaxants such as midazolam have been used to this end where there is spasticity and hypertonia of the TMJ muscles.[22] The greatest challenge is where none of these treatment options provide a solution. In these cases use of antimicrobial gel and/or aqueous lubricating agents, peri- and intraorally, provide some relief and aid prevention of infection.[22] Predisposing factors such as posture and positioning, patient comfort, illness or presence of pain, should be minimised where possible and Bobath techniques have been shown to be effective.[19,22]

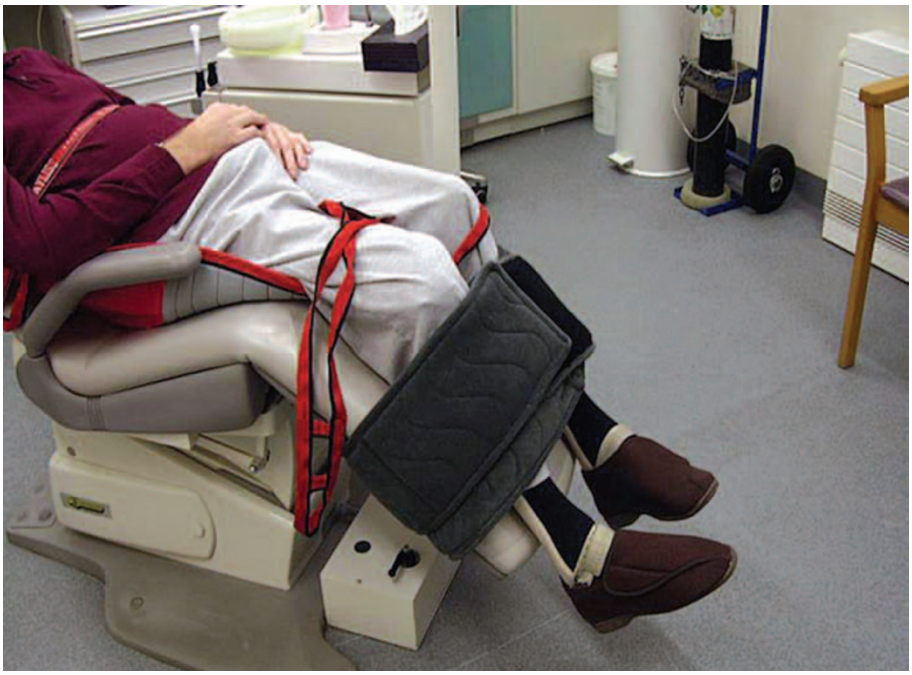

Fig. 6 Use of gentle restraint of legs with wide Velcro strap to control ataxic or jerky leg movements

Dental treatment

The provision of dental treatment can present its own set of challenges. However, with knowledge and understanding of CP, an empathic approach and careful planning, they can mainly be overcome. They include:

Consent – this should be assessed on an individual basis and it is essential that assumptions about capacity are not made on the basis of appearance or speech, which can be misleading.[1] Article 3 in this series provides further information on this topic.[33]

Access – some individuals with CP are wheelchair users and may require assistance or transfer aids, such as a hoist (Fig. 4) to access the practice or the dental chair. This has been covered in detail in article 1 of this series.[32]

Preventive dentistry – counselling about diet, oral hygiene and the use of fluorides is important for this group of people.[1] Suitable toothbrush adaptations (as outlined in article 4 of the series) may be crucial to achieve optimal self-care and liaison with carers will be required where they are responsible for providing oral care.[24] The use of sprays and gels in place of mouth-rinses is more appropriate for individuals who have dysphagia and poor oromuscular co-ordination.[34]

Communication – may be compromised by speech impairment and sensory deficits and require the use of communication strategies such as the use of hearing loops, computer assisted technology and AAC aids (Fig. 5).[3,32,35] Article 2 in this series considers communication in detail.[35]

Operative procedures – require careful consideration of physical posture and positioning of the subject, through appropriate support with cushions, etc, during treatment.[22] Together with use of high volume suction and/or the use of rubber dam, it will aid prevention of choking or silent aspiration during treatment due to swallowing difficulties and poor protective airway reflexes.[4] Sudden movements may require the use of physical intervention, with the patients consent, such as the gentle restraining of a limb[36] (Fig. 6), or the use of a mouth prop or finger guard to prevent sudden closing due to muscle spasm or the bite reflex.[1] Restoration of fractured anterior teeth is important to prevent discomfort and, more so, to restore self-esteem and social acceptability. For the latter two reasons, it can be argued that aesthetics is of greater importance to the person with disability than it is for the individual with no disability,[37] and a full range of restorative treatment should be made

available to them. There are no contra-indications to the use of local analgesic agents for restorative care or surgery, although access to sites of some injections maybe limited (Fig. 7).[1]

Anxiety and movement management – anxiety may worsen the usual athetoid movements or spasticity, and anxiolytics or muscle relaxants may be useful as a pre-medication.[1] Oral sedation may be effective but its effectiveness is unpredictable in patients who are taking other neuroepileptic drugs.[37] Article 5 of this series provides further information.[38] Inhalation sedation is an excellent option, however the requirements of continuous nasal breathing may be difficult for individuals with some physical disabilities or intellectual disability.[37] Manley demonstrated that the use of conscious sedation provides a valuable solution to providing a good standard of care in patients with CP. It enables repeated treatment sessions, often in primary care, and thereby opens a full range of treatment options.[37] He advocates the technique of titration of intravenous midazolam against patient response, with the suggested use of 0.25 ml of 40 mg/ml intra-nasal midazolam initially if required to overcome difficulties with cannulation due to movement disorder, patient co-operation, and/or compromised cognition. For people with profound disabilities or movement disorders, some care may need to be completed under GA due to poor co-operation or to ensure airway protection. This treatment modality can limit treatment provision if they are lengthy procedures or require repeat appointments.[37] Nor is it without risks in this group, due to the propensity for gastro-oesophageal reflux, poor pharyngeal reflexes, liability to aspirate material into the lungs and increased risk of hypothermia.[1,7]

The preventive and dental needs of most people with cerebral palsy can be met in general dental practice. However, some people, mainly those with profound neuro-disability, anxiety or learning disability, require either a shared approach to care or referral for specialist care.

2. DOWN'S SYNDROME

Down's syndrome (DS) is a genetic condition caused by a chromosomal abnormality (usually trisomy of chromosome

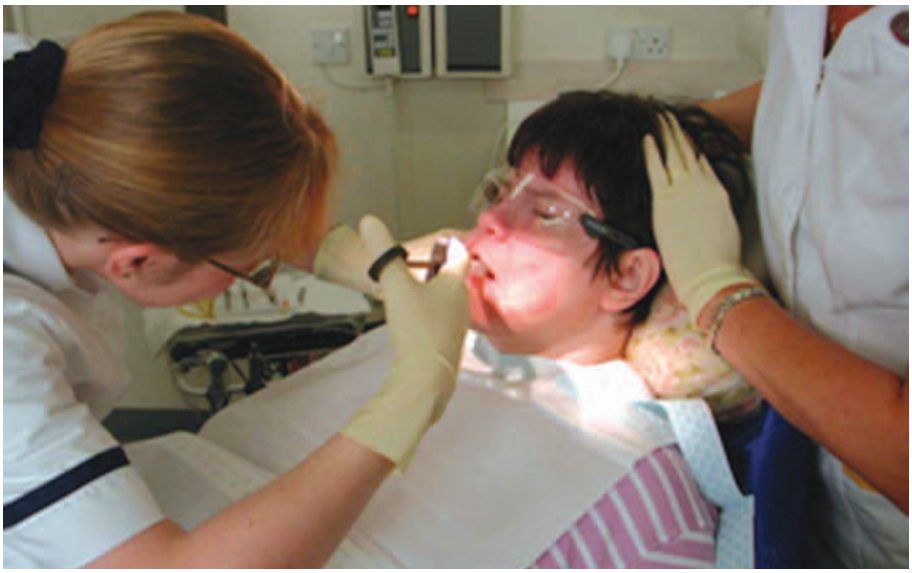

Fig. 7 Use of head support to control possible movement during administration of a local anaesthetic

Fig. 8 The characteristic facial features associated with Down's syndrome

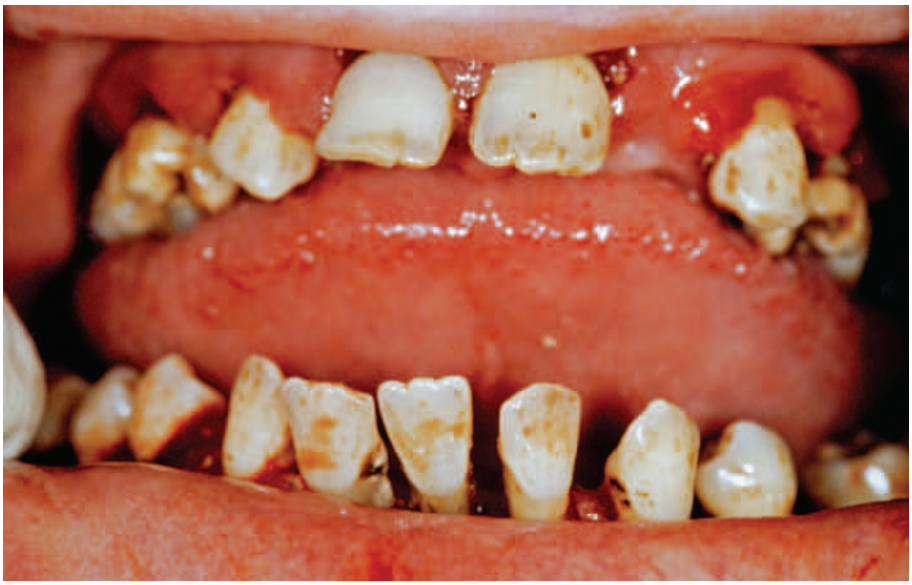

Fig. 9 The characteristic dental features associated with Down's syndrome

21) that results in a characteristic appearance, learning disability (which ranges from mild to severe) and a variety of physical and medical features.[39]

The characteristic appearance in DS is that of short stature, relatively short arms and legs, broad hands and short fingers, flattened face and occiput, slanting eyes with prominent epicanthic folds and underdevelopment of the middle third of face resulting in relative prognathism (Fig. 8).

Oral and dental characteristics include delayed development and eruption of both dentitions, hypodontia, microdontia, short roots, hypocalcification and hypoplastic defects, occlusal problems, and a high incidence of severe early onset periodontal disease (Fig. 9).[40] Physical and medical features include cardiac anomalies (40%),[41] visual impairment (50%),[42] hearing impairment (mild to moderate in 50%),[43] atlantoaxial instability or subluxation (20%),[44] compromised immune system,[41] hypothyroidism (15%),[45] increased risk of epilepsy (2-10% depending on age),[46] increased risk of diabetes Type 1 (2%)[47]and earlier onset of Alzheimer's disease.[48] All of these conditions need to be considered when providing dental treatment.[49]

Living longer

There are currently more than 26,000 people with DS in the UK and an incidence of 1 in 1,000 births, both male and female.[46] The life expectancy of people with DS has improved dramatically from an average of nine years in 1900 to an average of 50-60 years currently. As the prevalence of DS is set to rise, ageing in DS is only beginning to be researched and addressed.[50] Although many people with DS are able to live healthy adult lives without concerns related to serious illness or additional disability, it is reported that the health needs of older people with DS are not yet being met and some people with DS are dying from manageable and treatable conditions.[51]

Although life expectancy has increased, it is still lower than for the general population. Possible explanations are that people with DS 'age prematurely' and thus life expectancy is reduced; and that DS is associated with an increased risk of illness and mortality. The former explanation is generally favoured, and it is thought that the ageing process starts sooner or is speeded up. Subtle memory losses, physical tiredness and general frailty, as well as specific illnesses, may be present when a person with DS is in his/her thirties rather than his/her sixties. However, Alzheimer's disease (AD) is the only condition associated with decreased life expectancy that occurs earlier in DS.[51] It increases from about the age of 30, and by their fifties around 50% of people with DS show signs of AD. It increases with age at a similar rate as in the general population, but 30 or 40 years sooner. Its onset can be difficult to detect as in people with DS it may affect personality or behaviour before the classical early features of memory loss become apparent.[48] Dementia will be explained in more detail in the next article in this series. Diagnosis is based on the exclusion of other conditions that might present with similar symptoms (Table 2).

People with DS experience the same conditions as the general population as they age but experience them at an earlier age. However, they may be misinterpreted and where, for example, hearing or visual impairments lead to a decline in communication or living skills, they can be misdiagnosed as depression.[48] Depression itself is one of the most frequently diagnosed psychiatric disorders in the DS population, but is probably under-reported as people with DS may find it difficult to express how they are feeling, complicating its diagnosis.

Oral health

Despite an ageing DS population, most of the literature about oral health and DS relates to children and adolescents.[52-55] These studies generally indicate a lower caries rate than in the child population as a whole. However, Davila *et al.* found 53% of their study population had caries[56] and a study of 20-40 year old adults with learning disability living in an institution found a significantly lower DMFT in people with DS compared with those with cerebral palsy or idiopathic developmental delay.[57] A Hong Kong survey looking at the oral health status of 65 community dwelling adults with DS aged between 17 and 42 years found a lower caries rate in the DS group than the matched control group. The adults with DS had significantly fewer filled teeth, fewer decayed teeth, more peg-shaped maxillary lateral incisors, and more retained deciduous teeth.[58]

Table 2 Differential diagnosis of behavioural and functional change in later life in people with Down's syndrome

Depression
Hypothyroidism
Sensory impairments, visual and/or hearing
Dementia, usually Alzheimer's disease
Impact of major life events, eg bereavement
Other rare illnesses

Source: reference 48

The low caries prevalence in children with DS has been linked to immune protection from elevated salivary *Streptococcus mutans* IgA concentrations.[59] However, a study among 39 people with DS aged from 11 to 69 years demonstrated a lower rate of salivary secretion in people with DS than in a non-DS control group. It was attributed to decreased stimulated parotid salivary flow and, although not statistically significant, decreased with increasing age. Thus it is possible that caries may become more of a problem as people with DS age, although as yet there is no evidence to support this hypothesis.

The severity, prevalence and extent of periodontal disease are all significantly greater in the DS population than in the general population.[60] Prevalence has been reported from 58-96% for people with DS under the age of 35 years,[60] with lower incisors and upper first molars most commonly affected.[61] This situation is not attributed solely to poor oral hygiene and there has been a focus on an altered immune response due to the underlying genetic disorder of DS.[60] Findings regarding the management of periodontal disease in people with DS are mixed. Zigmond *et al.*[61] reported that a preventive programme had no effect on reducing the progression of either generalised or localised periodontal disease, indicating that impaired oral hygiene plays a relatively minor role in its pathogenesis. This is at odds with

the findings of Zaldivar-Chiapa *et al*., who reported that while there is partial impairment of immunological functions in people with DS, this did not seem to affect the clinical response to surgical or non-surgical periodontal therapy in a group of 14 people with DS aged 14 to 30 years.[62] This is supported by the work of Yoshihara *et al*., whose results suggest that periodic preventive care (at one to three month intervals) is effective in suppressing the progression of periodontal disease in young adults with DS aged 15 to 26 years.[63] Positive findings are also reported by Cheng, Leung and Corbett,[58] who achieved satisfactory healing responses following non-surgical periodontal therapy with the adjunctive use of chlorhexidine and monthly recalls in 21 adults with DS aged 20 to 30 years.[58] While the balance seems to be in favour of preventive programmes improving the periodontal situation for people with DS, the programmes require a degree of intensity and/or monthly review.[58,63,64]

The short roots of teeth in DS combined with increased periodontal disease make it probable that tooth loss from periodontal disease is more likely. There would seem to be no evidence in the literature to support or refute this supposition. The limited information relating to tooth wear is in the child population only and the findings are mixed. Bell *et al*. report it as significantly more common than in the general population (59% and 8% respectively),[65] with an aetiology of attrition and erosion, while more recently, bruxism has been reported as no more common in children with DS than in those without it.[66,67] There appears to be no literature indicating whether tooth wear is a problem in adults with DS.

Seamless care

The majority of dental treatment for most people with DS should be possible in the primary dental care service. Achieving patient co-operation is based on building trust and rapport through the use of behavioural management techniques such as acclimatisation and 'tell-show-do'. The degree to which this is successful in people with a learning disability may depend on the severity of the learning disability. However, once co-operation is established, local analgesia is the first line of treatment for most dental procedures for most people.[69] The choice of technique must always take account of any systemic disease, such as congenital heart and neurological conditions. Shared care with specialist support can be put in place if conscious sedation or general anaesthesia is required.

Access to dental care is essential for adults with DS in order that a rigorous preventive regime that will hopefully control periodontal disease and reduce the risk of tooth loss can be provided. The literature suggests that this should include daily adjunctive chlorhexidine and professional input on a monthly basis. Tooth replacement is not straightforward for this group of people. Dentures are difficult, although not always impossible, for people with learning disability to manage. Attention needs to be given to denture design – making it as simple as possible, avoiding gingival margin coverage and providing as good retention as is possible. Patience is required on the part of the dentist and the individual with DS. The use of adhesive bridges can be compromised by small crown size and/or spacing between teeth. While the literature related to the use of dental implants for people with DS is sparse, the two papers available suggest that implant dentistry is a viable treatment option[70,71] provided there is support from carers for the provision of good oral hygiene.[71]

In dealing with adults with DS in their forties and fifties, it must be remembered that some of them will have elderly parents who may find it increasingly difficult to support them in their oral hygiene needs. They will need information, advice and support to maintain their motivation. Many of the issues associated with seamless care for middle-aged people with DS have been explored in greater depth earlier in this series of articles, for example issues related to physical access to the surgery in article 1,[32] communication in article 2,[35] capacity and consent (including physical intervention) in article 3[33] and provision of information and materials related to oral hygiene in article 4.[24] The reader is referred to them for further information.

The dental team are important members of the DS multidisciplinary care team, as a healthy mouth can reduce the problems associated with DS and help to maintain the individual's self-esteem, quality of life and social acceptability.

3. CARDIOVASCULAR DISEASE

Cardiovascular disease (CVD) is the most common cause of adult death in the developed world. Dental procedures and drugs used in dentistry can aggravate heart disease and it is important that the dental team are aware of the common cardiac conditions and their management, as well as how to best manage the oral care of this group. The risk factors for CVD are shown in Table 3. Although precise mechanisms of interaction remain unclear, sufficient evidence exists to conclude that periodontitis places certain patients at increased risk of developing CVD.[72-74] Dentists need to take a careful medical history to ascertain the patient at risk of CVD.

a) Hypertension

Hypertension is a persistently raised blood pressure >140/90 mm Hg. Ninety percent of cases are 'essential', with no obvious cause, although smoking, diet and lifestyle are recognised causes. Pharmacological intervention should be offered to patients with persistently high blood pressure of over 160/100 mm Hg, with the aim of maintaining it at or below 140/90 mm Hg to reduce the risk of cardiovascular disease and death.[75] Antihypertensive drug management includes the use of diuretics, beta blockers, calcium channel blockers, ACE inhibitors, sympatholytics and vasodilators.[76] A significant number of people are in receipt of anti-hypertensive therapy, with up to 5% and 13% of patients attending general dental practice and dental hospitals, respectively, reported to take anti-hypertensive drugs.[77] Stress, including that associated with dental treatment, may further increase an already raised blood pressure, leading to a risk of stroke or cardiac arrest.[78]

The National Institute for Health and Clinical Excellence (NICE) recommends patient-centred care for management of hypertension, taking account of individual needs and preferences and providing

Table 3 The risk factors for cardiovascular disease
Risk factors for cardiovascular disease include:
• Smoking
• Excess alcohol
• Diabetes mellitus
• Hypercholesterolaemia
• Family history of cardiovascular disease
• Sedentary lifestyle
• Obesity
Source: reference 78

evidence-based information to allow patients to reach informed decisions about their care.

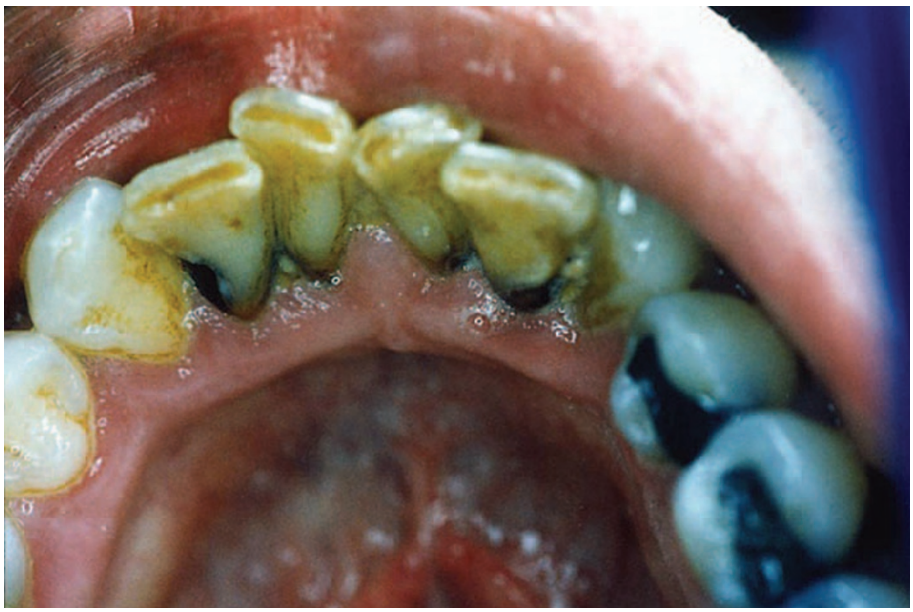

Fig. 10 Localised lingual caries associated with prolonged use of GTN tablets

b) Angina

Angina is severe, crushing chest pain. Stable angina is typically precipitated by effort and relieved by rest within ten minutes. The usual cause is coronary atherosclerosis resulting in insufficient blood flow to and oxygenation of the heart muscle.[78] The pain typically occurs behind the sternum, radiating to the left upper arm and occasionally to the left mandible, and rarely to the teeth, tongue or palate. Unstable angina is that occurring at rest, on minimal exertion or with rapidly increasing severity. Both forms are relieved by sublingual glyceryl trinitrate (GTN) spray or tablets. Unstable angina carries a significant risk of myocardial infarct and elective dental treatment should not be carried out. Surgical treatment using either stents or coronary artery bypass grafts has a good survival rate.

c) Myocardial infarction

Signs and symptoms of myocardial infarction (MI) are similar to angina but are more severe, of longer duration and are not relieved by GTN. The dental team should be aware that some myocardial infarctions are 'silent' and occur without any recognised symptoms or signs at the time.

Seamless care

For patients with hypertension, preventive advice and information on access to oral healthcare should be instituted at diagnosis to avoid the need for dental care later on. If feasible, dental treatment is best carried out using local analgesia, with or without conscious sedation. Anxiolytic agents and use of sedation are valuable tools for reducing the effects of stress while maintaining oxygenation and obviating the need for general anaesthesia. Side-effects of beta blockers can include xerostomia and appropriate management of dry mouth needs to be instigated.[24] Also, calcium channel blockers, particularly nifedipine, have been associated with gingival overgrowth which is best managed through good oral hygiene but may require surgery.[76,79]

Both beta blockers and non-potassium sparing diuretics can exacerbate the effects of epinephrine in dental local anaesthetic agents and it is recommended that patients with mild to moderate CVD receive the smallest amount of local anaesthetic needed to provide effective analgesia, using an aspiration technique to prevent intravascular injection.[80] Many patients with CVD may be taking anticoagulants such as aspirin or warfarin and the management of these individuals is described in article 5 of this series.[38]

Effective analgesia, short appointments and availability of both oxygen and GTN are all important in treatment regimens. Prophylactic GTN prior to dental treatment has been shown to be effective in the prevention of both hypertension and angina during treatment.[81] GTN should also be easily to hand throughout dental treatment and should relieve chest pain in angina within five minutes. Prolonged use of GTN tablets has been found to cause caries localised to the area where the tablet is retained (Fig. 10).[82] This can be avoided by using a GTN spray.

It is commonly recommended that patients do not receive dental care for at least six months after experiencing an MI.[83,84] However, Meechan suggests that ideally, elective treatment should be postponed for a year as there is a high chance of a further infarct during this period.[78] Until this time, acute dental needs should be managed in consultation with the patient's physician. All patients with CVD should be managed using a stress-reduction protocol that includes short appointments, preferably in the morning when patients are well rested; use of effective local anaesthetic to minimise discomfort; use of conscious sedation to reduce stress; and provision of excellent post-operative analgesia.

d) Congenital cardiac conditions and acquired cardiac disease

Infective endocarditis (IE), although uncommon, may affect damaged heart valves, prosthetic heart valves, a coartated aorta, patent ductus arteiosus or ventricular septal defect. As *Streptococcus viridans* is the most commonly isolated bacteria in IE,[78] until recently prophylactic use of antibiotics was

recommended for dental treatment in patients with acquired valvular heart disease with stenosis or regurgitation, valve replacement, structural congenital heart disease, previous IE and cardiac myopathy. Although these groups are still considered to be at risk of developing endocarditis, after reviewing the literature, recent NICE guidance has removed the need for antibiotic cover for dental treatment as no evidence could be found that this regime prevents infective endocarditis.[85] Patients who have received antibiotic cover for dental treatment for many years will require careful counselling to help them understand the situation and to avoid confusion and distress.

Seamless care

NICE recognises the pivotal role that both the individual and the dental professional play in maintaining oral health.[86] It is paramount that this group of patients have ready access to oral care and they have every opportunity to achieve and maintain good oral health in the primary dental care setting. The responsibility of the dental team is to educate the patient about, and to alert them to, the signs of infective endocarditis (Table 4) and when to seek expert advice; and to instil in them the importance of good oral hygiene and regular reviews and dental treatment as appropriate.

Table 4 Indications of infective endocarditis

Systemic signs	Low grade fever and generalised malaise develops and persists Weakness Arthralgia Loss of weight Sepsis of unknown origin
Skin	Pallor (anaemia) Light pigmentation (*café au lait*)
Embolic phenomena	Purpura of skin and mucous membranes Splinter haemorrhages under fingernails Janeway lesions (small painless erythematous or haemorragic stigmata in the palms of hands or the soles of feet Oslers nodes (tender vasculitic raised lesions in the skin, usually on the digits) Haematuria Stroke or transient ischaemic attack
Cardiac	Changing or evolving cardiac murmurs Loss of peripheral pulse

Source: reference 100

e) Inherited and acquired bleeding disorders

This subject has been dealt with in article 5 of this series and the reader is referred to it for information.[38]

Given the high mortality rate of CVD and the relatively minor morbidity of periodontal therapy, maintenance of periodontal health should be among the routine recommendations for prevention of heart disease.[87]

4. RESPIRATORY DISEASE

This section covers only the respiratory diseases most likely to be encountered in dental practice, namely chronic obstructive pulmonary disease and asthma.

a) Chronic obstructive pulmonary disease (COPD)

Chronic obstructive pulmonary disease (COPD) is the name for a collection of lung diseases including chronic bronchitis, emphysema and chronic obstructive airways disease, all of which can occur together. These diseases have different aetiologies but may have overlapping signs and symptoms.

COPD is the most common chronic condition in the UK, varying in severity from mild through to disabling and severe disease with respiratory failure.[88] Most commonly it affects people over the age of 40 and it is thought that around 900,000 people in England and Wales have been diagnosed with COPD, although the true number may be around 1.5 million.[89] It is predominantly caused by smoking and is almost entirely preventable.[90] It is characterised by airflow limitation that is not fully reversible, is usually progressive, although it does not change markedly over several months, and is due to a combination of airway and parenchymal damage resulting from chronic inflammation. Other factors, particularly occupational exposures, may also contribute to the development of COPD.

Many dentists will remember learning about 'pink puffers' and 'blue bloaters'. The former are patients with emphysema who maintain normal blood gases by hyperventilation and are breathless but not cyanosed, while the latter are patients with chronic bronchitis who fail to maintain adequate ventilation, becoming hypercapnic and hypoxic and developing central cyanosis, cor pulmonale and oedema.[91] Cor pulmonale or pulmonary hypertension relates to the heart being affected adversely as a result of raised blood pressure in the lungs and may occur as a direct result of COPD.

There is no single diagnostic test for COPD and diagnosis relies on clinical judgement based on a combination of history, physical examination and confirmation of the presence of airflow obstruction using spirometry.[90] The treatment of the disease is tailored to the severity of the symptoms and the cornerstones are smoking cessation, inhaled bronchodilators and inhaled corticosteroids. Patients with severe COPD may need nebulised oxygen at home for up to 15 hours a day. This can improve shortness of breath but limits activities and may mean they are confined to home. The impact of COPD on the quality of life of individuals and carers can be considerable[92] and they are also prone to anxiety and depression.

Seamless care

Patients with COPD should have access to the wide range of skills available from a multidisciplinary team (MDT). Professionals involved in their care may include a respiratory nurse specialist, specialist physiotherapist, occupational therapist to assist with managing daily living activities and, for people with end stage disease, the palliative care team. The

dietician may also be involved, as about one third of people with COPD have significant malnutrition related to diminished appetite and the increased energy expenditure required for breathing.

The dental professional can be a valuable member of the MDT as there are a number of oral health risk factors. The frequency and severity of periodontal disease is increased in people with COPD.[93] The oral health risk can be exacerbated if the individual is a smoker, or if they are on oxygen therapy, which is associated with xerostomia. Additionally, patients may be advised to eat small, nutrient and calorie-rich meals frequently. The dental team have a role to play in the provision of oral health advice, smoking cessation and dietary advice. This is particularly important, as poor oral hygiene has been linked to respiratory pathogen colonisation and dental plaque may act as a reservoir for acquired pneumonia in older people, particularly residents of long-term care facilities.[94]

Most people with COPD can receive dental treatment safely, with only minor adjustments to procedures, in general dental practice. For comfort of breathing, they may need to have their dental treatment in an upright or semi-reclined position. People on oxygen therapy will need ambulance transport for dental appointments and an adequate supply of oxygen during the visit, or provision of domiciliary oral healthcare. Patients with severe COPD are at particular risk when given intravenous sedatives, opiates or general anaesthetics due to respiratory depression, and where possible should be treated with local analgesia.[88] The respiratory centre of a 'blue bloater' is relatively insensitive to carbon dioxide and the individual relies on hypoxic drive to maintain respiratory effort. Providing supplemental oxygen for more than brief periods can be dangerous to such patients as without the hypoxic drive they may hypoventilate or stop breathing altogether.[95]

b) Asthma

Asthma is common. It is estimated that 5.2 million people are affected in the UK, with at least one affected person in every five households.[96] In 2005, 1,318 UK deaths were attributable to asthma. It is described as a generalised airway obstruction which, in the early stages, is paroxysmal and reversible. The obstruction is due to bronchial muscle contraction, mucosal swelling and increased mucous production and leads to coughing, wheezing, and/or shortness of breath.[96] Common triggers include house dust mites, animal fur, pollen, tobacco smoke, cold air, chest infections and stress, and adult asthma can develop after a viral infection. There is no cure for asthma and treatment and management include the use of preventer and reliever inhalers. If this proves insufficient, inhaled steroids are used and in severe cases systemic steroids may be prescribed.

Seamless care

Anti-asthmatic medication, such as salbutamol inhaler or tablets and beclamethasone inhaler, can lead to both increased dental caries and periodontal disease. In order to control any possible exacerbation of dental disease, people with asthma should be advised by their doctor, pharmacist or dentist that they need to adopt more precautionary oral hygiene practices and have regular dental reviews.[97] Salbutamol is a ß2 adrenergic agonist and can produce dry mouth, taste alteration and discolouration of teeth. People using corticosteroid inhalers are also predisposed to developing candidosis.[98,99] To help prevent these side-effects, people are advised to rinse and gargle with water and brush their teeth after using their preventer inhaler.[96]

Before dental treatment, an asthma history that includes efficacy of medication, use of steroids and any episodes of hospitalisation should be ascertained.[95] If steroids have been taken long-term, consideration should be given to the need for increasing the dose prior to invasive dental treatment. The severity of an individual's asthma will vary, so it is prudent to plan dental care around periods when the condition is less severe. As exposure to allergens and/or stress can induce an asthma attack, it is advisable for the individual to use their inhaler prior to treatment and to have it available throughout the appointment. Additionally, efforts should be made to allay anxiety as far as is possible. Preferably, treatment should be carried out with local analgesia.[95] If conscious sedation is required, relative analgesia is the technique of choice as in the event of an asthma attack, it can be more rapidly controlled than intravenous sedation.[95] Aspirin is not recommended for analgesia as many people with asthma are allergic to it.[95] Also, use of non-steroidal anti-inflammatory drugs (NSAIDs) may precipitate an asthma attack and it is safer to recommend the use of paracetamol.

If an asthma attack occurs on the dental premises, the individual should use their reliever inhaler immediately, sit but not lie down, and loosen any tight clothing. If there is no immediate improvement, they should continue to use their reliever inhaler at the rate of one puff every minute for five minutes or until symptoms improve. If symptoms do not improve in five minutes the emergency services should be called and the reliever inhaler use continued every minute until help arrives.[96]

Although a large proportion of people with respiratory disease are able to receive routine dental treatment in general dental practice, those with significant respiratory problems are best treated in a hospital setting.

The illustrative material used in Figures 1, 2, 3 and 5 is credited to www.JohnBirdsall.co.uk. The authors would like to thank the Down's Syndrome Association for providing them with the illustrative material used in Figure 8.

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