RETT SYNDROME ASSOCIATION UK

113 Friern Barnet Road, London N11 3EU
Tel: 0870 770 3266 Fax: 0870 770 3265
Local Callers Tel: 020 8361 5161 Fax 020 8368 6123
E-mail: info@rettsyndrome.org.uk
www.rettsyndrome.org.uk

Dental Health for People with Rett Syndrome

Mary Henderson, B.Sc., B.D.S., M.Sc.
Assistant Clinical Director/ Senior Dental Officer Special Care Dentistry
Specialist in Paediatric Dentistry/ Eastern and Coastal Kent PCT Dental Service

Dental Problems and Rett Syndrome

Parents and carers of individuals with Rett syndrome often do not seek dental care early on, e.g. as a baby, toddler, or young child, as it is about this time that problems with feeding, mobility, and communication are noted,[1] and as a continuous round of visits to professionals may be occurring whilst a diagnosis is being made.

Often the first visit to the dentist is for emergency care or pain relief, which can be a difficult time for both carer and patient. It is important to try to find a dentist who has time and understanding; it may be useful to have a dentist experienced in the care of special needs patients, and the best time to find a suitable dentist is when no problems are occurring. This enables the patient to become used to being seen by the dentist and familiar with the environment, and preventive therapy can be provided.

Patients with Rett syndrome have the same oral and dental problems as the rest of the population, and can suffer from tooth decay and gum disease. Both can lead to pain, suffering, and the possibility of tooth loss. Both diseases are preventable, but some of the problems associated with Rett syndrome make dental management more difficult. Prevention for this group is a priority, as dental treatment may require more complex or invasive ways of providing treatments, such as sedation or general anaesthetic.

I. Preventing Tooth Decay

Tooth decay only occurs if teeth are exposed over time to foods and drinks that are high in sugar. Restricting the number of times a day sugar is consumed is the most important way of preventing tooth decay. If sugar is consumed, it should be as part of a meal, e.g. at the end of the meal as a sweet.

Water and milk are considered safe drinks, but drinks with sugar will rapidly decay teeth, e.g. blackcurrant juice. Squashes should be sugar-free; any acidic drinks should be well diluted and used infrequently. If used often, especially in a bottle, they will rapidly affect the front teeth first.

Chewy foods that stick in the fissures or cracks of teeth such as biscuits, sweets, or sugared breakfast cereals should be avoided. These will decay the back teeth first; and later will affect the whole mouth.

Sugar-free drinks and even sugar-free sweets can help reduce the sugar content of the diet. Sugar-free sweets contain a molar symbol with an umbrella above, representing

“Tooth-friendly” sweets. It is best in the long term to encourage a diet of healthy foods that include fruits and vegetables rather than substitution of one junk food for a “healthier” alternative.
Medicines are often prescribed to children in a syrup or suspension. They are often given last thing at night, when the protective salivary flow decreases. Most medicines can be obtained in a sugar-free formulation, but often you must ask the prescribing doctor or, if purchased over the counter, the chemist.

II. Improving Resistance to Tooth Decay

A. Sugar and Diet

Resistance to tooth decay can be increased by the use of fluoride. We are aware that fluoride becomes incorporated into tooth enamel by being close to the tooth surface. Thus, fluoride toothpaste is a good way to raise resistance of teeth. In a child, generally under 6 years of age, who will not spit our toothpaste, a small pea size amount of children’s toothpaste should be applied to the toothbrush and scrubbed into the teeth by parents or carers at least twice daily. Children’s toothpastes generally contain a lower dose, 500 ppm of fluoride to reduce the risks of mottling (white and brown patches) in forming tooth enamel. If a child of 6 years is able to spit, then adult toothpaste can be used, but a pea size is adequate.

B. Fluoride

Fluoride supplements can be administered in drop or tablet form, as prescribed by your dentist; these allow some fluoride to become incorporated into the developing permanent teeth, as well as exerting an effect as the surface of the tooth which reduces decay. These are best taken on the advice of a dentist who will consider the level of risk of decay for the child and the fluoride levels in the water, which can vary from area to area (and well to well). The local Water Board will know the concentration of fluoride, if any, in the local drinking water for a given address.

Fluoride varnish can be painted on the teeth by the dentist to prevent decay occurring. Usually the dentist will target fluoride application to areas that are prone to decay.

C. Fissure sealants

To improve resistance to decay beginning in the cracks (pits and fissures) in molar teeth a thin coating in plastic can be painted on the molar teeth if cooperation allows. These are known to reduce decay by approximately half in pits and fissures, but they do require the tooth to be kept dry when the coating is applied.

III. Healthy Gums

Plaque deposits (the yellow, white, furry bacteria that grow on the teeth) accumulate, causing the gums to be red, swollen, tender and enlarged. This usually happens in the area where the gum meets the teeth. If this inflammation is not controlled, the whole bony support for the teeth can be lost; gums recede, teeth loosen and are eventually lost due to periodontal (gum) disease.

Patients should have their teeth brushed by their parent or carer using a soft to medium nylon-bristled toothbrush. Electric toothbrushes are useful where patients are able to brush their own teeth but lack the manual dexterity; most will need supervision and carers

brushing for them. Coming from behind, with the individual sitting or lying down is the most efficient way to brush a person's teeth. Use of chlorhexidine gluconate in the form of gel, mouthwash brushed on the teeth, or spray are available. Advice and guidance from a dental professional such as a dentist or hygienist should be sought before use of chlorhexidine.

Bleeding gums need to be kept clean in order to reverse the inflammation (gingivitis). Healthy gums should not bleed; if bleeding occurs, it can usually be reversed with good tooth brushing.

IV. Factors in Rett Syndrome Influencing Dental Treatment

A. Epilepsy

Teeth should be checked regularly as they may fracture or even be knocked out when falling occurs.

Some medicines used to control epilepsy e.g. Phenytoin, can cause overgrowth of the gums. Plaque removal is even more important for patients who are at risk of this overgrowth; plaque can accumulate even more when gums are swollen.

B. Medicines and Drugs

Be aware of the sugar content of medicines. Ask for sugar-free formulations at the doctor and at the chemist's for over-the-counter medicines. If sugared medicines must be used, try to avoid giving them last thing at night.

Some drugs can cause a dry mouth, which can increase the risks of tooth decay. For patients who need these drugs, increased use of preventive measures such as use of fluoride may be appropriate. It is important that patients have plenty to drink, especially if they drool, to avoid dehydration.

C. Drooling and Dribbling

This can occur because of posture, poor oral seal and infrequent swallowing, and poor jaw or lip closure. Saliva is important for oral cleansing, to protect teeth from decaying, to provide lubrication for food and protect soft tissues, and it actually allows food to be tasted. Digestion begins within saliva; its enzymes begin to break down carbohydrates and protein.

If drooling is a problem, a multidisciplinary team including the dentist may be able to help. It is important that teeth are not lost to allow gaps for saliva to pour out where posture is poor.

Occasionally, in severe cases, drugs or even surgery are used to reduce salivary flow; if this does occur then prevention of tooth decay with use of increased tooth cleaning and fluoride is important; and dental checks may need to be more frequent.

D. Tooth Grinding (Bruxism)

This can be extremely frustrating for carers as it is noisy to listen to, and it ultimately wears away the teeth. Whilst worn teeth can look unsightly, the wear is often slow enough for the pulp (nerve) of the tooth to protect itself. In severe cases, plastic splints can be made, but

should be monitored carefully, as they can have stagnation around them with plaque growth causing tooth decay if prevention is not undertaken.

V. Signs and Symptoms of Dental Problems

Apparent pain, a change in behaviour or sleep pattern, increased drooling or dribbling, fingers or hand in the mouth, redness and or swelling of the cheek or floor of the mouth, or refusal to eat and drink are possible signs of dental problems. Discolouration of a tooth, chipping of a tooth, mobility, or unexpected tooth loss all need to be investigated by a dentist.

It has been noted that Rett syndrome patients can have supernumerary teeth (extra teeth or fused teeth). These may make cleaning more difficult and they can also delay eruption of adjacent teeth. [2,3]

VI. Finding a Dentist and Regular Visits

The majority of dentists work in general practice (e.g. family dentists). A few work as Community or Salaried Primary Care Dentists (some of whom work exclusively with special care patients and have experience of patients with a variety of medical conditions, including Rett Syndrome). Other dentists provide care in a hospital setting, though this is more usually of a surgical or orthodontic work (monitoring growth and straightening teeth). Whilst it is a good idea for patients to be seen with their families in general dental practice, the dentist may feel that he or she does not have the resources or experience to treat a patient with Rett syndrome. They may also not have the resources required, e.g. sedation facilities, or access to general anaesthetic provision. Community (Salaried) dentists with training and experience in Special Care Dentistry may be able to provide this care, and are experienced in prescribing when this type of care is needed. Good management and sedation make the need for general anaesthetic less frequent. How to gain access to Special Care Dentistry in your area can be clarified through your PCT.

Time during a dental visit is important; having methods of accessing the mouth demonstrated can be helpful. Use of mouth props, bound tongue depressors and toothbrushes all work to assist in opening a patient's mouth to brush teeth. Home care should be discussed; patients should be made to feel relaxed and welcome. Carers should be made to feel part of the team, for it is a team approach to dentistry that will enable good home care to be undertaken.

Regular visits to the dentist and good home care can prevent the need for invasive dental treatment. Preventive methods used appropriately should reduce pain and infection for this special group. [4,5]

References

1. Crawford, A., Dental health for people with Handicaps Including Rett syndrome Rett2/information/dental health
2. Henderson, M.E., unpublished work
3. Buccino, M.A., **www.kidscare.com**
4. The Scientific Basis of Dental Health Education, Health Education Authority, Mabledown Place, London, WC1H 9TX
5. Nunn, J., ed., Disability and Oral Care, International Association for Disability and Oral Health and FDI, 2000.

Updated May 2007