ECTODERMAL DYSPLASIA AND ORAL HEALTH

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If I were to paint a pen portrait of a child with Ectodermal Dysplasia it would be something like this – a boy, maybe with thin, wispy hair, not much in the way of eyebrows, a dry looking skin often with dark circles under the eyes and a gappy smile. Whilst the gappy smile is something dentists expect in a toddler and again in the early school years, for the older child it is a cause of social embarrassment. This is particularly in the turbulent years of adolescence but can be an issue, as you know only too well, at the stage when a child first starts school. The lack of teeth is just one aspect for children so affected but there are other oral and dental problems that they may face and we can talk about each in turn.

Prevention, prevention, prevention!
Mouth cleaning
For the child in whom teeth may be missing - either primary (milk teeth, deciduous teeth) or permanent teeth, preventing dental disease in the remainder is vital. In my experience, children with large numbers of missing teeth tend to have clean mouths. Whether that is because there are fewer teeth to clean or whether the gaps make it easier, I am not sure, though parents report that cleaning single teeth is actually quite difficult. Cleaning the mouth and teeth, regularly twice a day, really well, is vital. You need to find a method and brush that suits your child - but your dentist or hygienist will help with this; insist on it!

Generally, the smaller the head on the brush, the easier it is to get to the back of the mouth. For a child who does not want to cooperate with brushing, using a ‘Superbrush’ can be a help. These brushes have bristles on three sides so that as you move the brush along the teeth, it cleans the top and both sides of the tooth at the same time. A single tufted ‘interspace’ brush is good to get into small gaps though it needs some help to manage this.

Cleaning is best done after breakfast and certainly before bed. Whilst a child is asleep, there is much less saliva in the mouth. Saliva (spit) has a major protective role, neutralising the effects of harmful acids produced by food left around the mouth.

If that food is not cleaned off before bedtime, the decay cycle can start and go unchecked for hours because there is just less saliva in the mouth at this time.

Cleaning effectively is more important than brushing lots of times in a day; no child has the physical skill to use a toothbrush properly until they are around 6 or 7 years of age. They need help with brushing – maybe you brushing around all surfaces after they have had a go themselves. Your dentist or hygienist should advise on the best way to keep your child's mouth clean.

Toothpaste
Using the right toothpaste is important too. Children under 6 years generally should use a children’s formula toothpaste. After 6 years of age they can use the same toothpaste as the rest of the family, provided they like the taste. Encourage your child to spit out any excess toothpaste but not to rinse out vigorously. That way they get the most benefit from the toothpaste.

Extra fluorides
If the area you live in does not have a fluoridated water supply your dentist may advise you to give your child a daily fluoride supplement. Again, check
this with your dentist who will decide the best preventive plan for your child.

**Dry mouth**

Most children with ED have less saliva in their mouth. This makes their mouth feel dry, food is more difficult to chew and swallow, talking can be more difficult and the child is more likely to develop dental decay. Saliva helps to neutralise acids produced from food and drinks. If you have less of it you are more likely to develop decay with holes that get bigger very quickly. Your dentist will advise on ways around this, which in adults usually involves replacing the saliva that is not there or taking drugs to encourage the remaining saliva glands to produce more. At the same time, it is vital to be really careful about what your child eats, especially sweetened foods and drinks. It is when and how often such things are eaten or drunk that is important. Snacking on and off frequently through the day is the most damaging habit for teeth.

**Food and drink**

Dentists can be real killjoys when it comes to diet! However, you will also be keen to preserve any teeth that your child has. The cornerstone of successful prevention of dental decay is keeping intakes of sweetened food and drinks to a minimum. Ideally, anything sweet should be taken at a meal and the number of intakes of such foods and drinks limited to 5 per day. Try to keep sweets and chocolate, as well as fizzy and other soft drinks, as treats to be eaten/drank after a meal or on a ‘sweetie day’. Resist the pressure to give in to your child’s demands for biscuits and sweets and offer safer foods, encouraging your child to eat fruit and savoury snacks. For drinks, stick with milk, water, sugar-free, well diluted squash, and of course, unsweetened tea and coffee as they get older. Try to keep food and drink clear of bedtime by about one hour since overnight the protective effect of saliva is much less.

**Dental Care**

**Advice**

Any baby should be registered with a dentist – even before they have any teeth! This is a crucial time for getting into good dental habits and to put into practice the advice your dentist or hygienist gives you – for example, safe weaning foods, baby drinks, when to start cleaning teeth and with what. If your dentist thinks that fluoride supplements are needed, they should be started roundabout the time that the teeth come through into the mouth. Your local health clinic may have a community dentist working there or will know where the nearest community clinic is if you do not have your own dentist. Sometimes these clinics specialise in children’s dentistry.

**Care Planning**

Ideally all children with ED should be seen by a multi-professional team (often called a Hypodontia Clinic) who have experience of working with children that have missing teeth. The team will usually include a paediatric dentist (somebody specialising in children’s dentistry), an orthodontist (someone who monitors growth and development as well as moving teeth with braces), a restorative dentist (someone who provides fillings, crowns, dentures, bridges and implants), an oral surgeon (a person who removes or uncoveres buried teeth, transplants teeth and carries out other surgical procedures) and a nurse coordinator who organises clinics and is available at the end of a phone on a daily basis, for any questions a family may have. It is vital that planning, both short and long-term, is done jointly with all these people who can together decide the most workable plan for you and your child.

**Treatment**

Children with ED may have fewer teeth than children of the same age and the ones that are present are sometimes pointed and small. This may be the case in the primary (milk, first) and/or permanent teeth. Your dentist will suggest a number of options to overcome some of these problems.

Small, pointed teeth can be made to look like ‘proper’ teeth by rounding-off with tooth-coloured filling materials and that goes for both primary and permanent teeth. Missing teeth can be replaced, if this is thought to be necessary by both the family and the dentist. For a child with primary teeth only, this is usually best done with removable plastic dentures (‘Plate’). Your dentist or a specialist in a community clinic or hospital will make these. Helping a child get used to these dentures is important and often the best person to do this is a family member or friend who already wears dentures and will be an expert on how to cope with false teeth. Most children adapt to these very quickly and often their school friends do not know the teeth are not their own.

For older children, joint planning at a multi-professional clinic including discussion with you and your child, may lead to an offer of orthodontic work followed by dentures (in the short term) and then bridges. When your child has finished growing, implants may be considered.

Waiting lists to be seen by multi-professional teams can be long so it is important that your dentist refers your child to a local team, where one exists, when s/he suspects that your child may have missing teeth. Where there is no multi-professional team much of this dental care will be carried out by an
orthodontist working with a restorative dentist in a regional centre.

Alongside all this, your child will need to see your own family dentist in order to have regular preventive care, for example, fluoride treatment and fissure sealants. Extra fluorides, as well as fluoride that your child will be getting from toothpaste, are used if your child is prone to decay. This is usually painted onto the teeth each time your child visits the clinic. The nooks and crannies (‘fissures’) on the biting surfaces of back teeth are difficult to clean. A toothbrush cannot reach to the bottom of the nooks and crannies and it is there that decay often starts.

Putting a sealant into the fissure (nooks and crannies) prevents germs lodging there and so prevents decay from starting. This is a painless, simple thing that your dentist or hygienist can do.

On a day-to-day basis, your local dentist will often help with minor problems like a sharp edge on a denture and, of course, even when your child is being seen by specialists, they should still visit their family or community dentist for check-ups and preventive care.

All dental care for children in the UK is free until a child is 18 or longer if in full-time education.

Are you a Teenager who has had Implants?

We have many youngsters embarking on the long road towards Implants. As a teenager who has had implants would you please write an article for our newsletter for the younger teenagers to read explaining such things as what it’s like, how long it takes, is it very painful, did you have to have bone grafted, what was that operation like, is it all worth it!

Disability Rights

We have, over the years, accumulated quite a lot of paperwork relating to ED and the various syndromes which may be of help to you when completing your Disability forms or attending Appeals. Please do not hesitate to contact me for help with this, the more paperwork you have to support your claim the more chance you have of obtaining it. At a recent Appeal it was stated that Ectodermal Dysplasia is not a recognised condition!! We must prove them wrong.

Hair Pieces

I am often asked questions relating to whether or not children from a very young age through to teenage years should, at any point, be offered hair pieces, what they are like, do the children wear them, where do you get them (the best place), does it help build their confidence, should we encourage or discourage them? If you have had any experiences would you please either contact Diana at the EDS office for a chat, or write an article for the newsletter?

Research

Wonderful news. The NFED have awarded John McGrath, a member of our Medical Advisory Board $24,000 (USD) for his research work on “Abnormalities of the p63 gene in ectodermal dysplasia and related syndromes”. John is working with two other members of our Medical Advisory Board and they are looking at the gene responsible for AEC (Haywells), Rapp-Hodgkin, EEC and similar types of ED. They have recently carried out a DNA based antenatal test for EEC syndrome, the first in this disorder and, as the test indicated, the child has been born unaffected. Most of the research uses genomic DNA from blood samples and all the work has been approved by local Ethics Committees.

The team are always looking for willing participants in their research programmes, so if you have AEC, EEC, RH or split hand/foot and are willing to help, then they will be very happy to examine the p63 gene for you. Please get in touch with the EDS office.
Incontinentia Pigmenti – small, but not insignificant

My daughter, Sarah, is 11 – she arrived 4 weeks prematurely, was footling breech presentation and had to be delivered by emergency caesarean section – a bit of a shock to all of us, including Sarah!

Whilst in hospital for the first week I had difficulty feeding her; the midwives said she was a very “windy” baby. Just prior to discharge a junior doctor saw her being “windy” and promptly rushed her to the Special Care Baby Unit – the “wind” was her having convulsions.

This was all a bit much to take – my little girl in an incubator covered in wires and on “phenobarbitone”.

However, the drugs stopped the convulsions* and we thought we’d be going home; but then the “nappy rash” that had been virtually ignored spread up her trunk and over her arms & legs – What now?!

*It’s thought that the fits were caused by her prematurity/delivery – not connected to IP.

Luckily for us, as I now know people have had to wait months or years for diagnosis, the attending Dermatologist thought immediately that it could be Incontinentia Pigmenti – asked for a biopsy and confirmed it. Although I said luckily – at the time we didn’t feel at all lucky. Nothing seemed to be known about this genetic condition. The paediatrician (a lovely man) went from books and could do no more than reassure us that the fits had stopped for now and tell us that Sarah would be carefully monitored. We got worried about all the things that might happen later – he didn’t know – nobody did.

She was carefully monitored, measured and assessed – but apart from a slightly larger head than normal (later dismissed as a family trait!) and a tendency for clumsiness everything seemed ok. The rash got worse, then cleared occasionally coming back if she was ill, but most things were just like all the other babies we knew.

The reason for her clumsiness was later found to be that she had no sight in her left eye (this hadn’t been a clear possibility in any of the literature that we had at the time) and was one of the biggest shocks since the fits. Sarah, though, didn’t know any different – they think she’s never been able to see from it, and got on with life very happily.

I didn’t know anyone else with IP until, when Sarah was about 5, I heard about Contact a Family through my Mum who was a health visitor at the time. I got in touch with them who put me touch with a couple of people (Moira and the ED group being one of them). Contact a Family have since given my number to a few people and we have had lovely chats and shared our experiences.

It was so nice to know that we weren’t the only ones – how I wished that these people had been contactable when I had Sarah. As it has turned out, apart from her eye, IP doesn’t affect Sarah too much – she’s just started Grammar School and enjoys life to the full. But if I’d had someone to talk to at the beginning, I’m sure I would have had far fewer sleepless nights.

IP, in general, isn’t particularly debilitating – but not knowing what lies ahead and feeling alone can be very debilitating for the parents.

Over the years I have spoken to a lot of people for long periods of time and then never heard from them again - which is great; knowing you’re not alone and that someone is at the end of a phone if necessary is often all that’s needed.

Having a link up for parents is also important so that the professionals have a point of contact for future parents, and so that any new research developments can be shared,

We may be a small group, but we must not be insignificant!

Alison Manley
Donations

Thanks to all who sent us warm Christmas wishes, and to those who enclosed stamps and donations. These are always greatly appreciated, and amounted to £115.

Translators

Is there anyone out there who speaks French or German, and would be able (and have the time) to translate some newsletters, 2 or 3 times a year?

Our counterparts in France and Germany each produce a newsletter which they send to me, and I can’t read them! Although I’m studying French, it will be a while before I can understand the technicalities in the newsletters. It would be great if someone could précis these so that I can keep abreast of developments in Europe. Please let Diana know at the EDS office.

Fundraising

We were grateful to receive £180 from the staff on the Winstanley Ward, Chorley & District General Hospital, who held a Christmas hamper raffle. Well done, and thank you for all the hard work you put into organising this event.

We were also pleased to receive £55 from the Coventry Fun Run, although at the time we did not know who had put our name forward! We now know – a huge thank you to Helen Wyer.

Do you like Running?

We are putting together a team of members/friends/family who enjoy running. We would like to hold a running event each year somewhere in the UK, together with the big events such as the London Marathon. If you would like to join this team, please let Diana know at the EDS office.

Anyone have any other ideas for fundraising?

Eye Operation

Elaine Stevens writes:

My eyes can be really troublesome at times because of the lack of oil glands. Even though I have plenty of tears in the daytime, winter nights with central heating and the lovely hot days, are not so enjoyable. With a little self-help with eye creams and eye drops, I must be a walking chemist!

Two years ago I had eye surgery, which was carried out at The Oxford Eye Hospital. I had surgery on my right eye because the skin on the eye was extra thick and cut the corner right off, making the eyesight poor. This condition has got worse over the last ten years.

Six months after the operation, the skin started to grow back again. I have now had two operations on this eye. The first time the surgeon cut the extra skin away and stitched the eye but it grew back within three weeks.

The second operation, with skin graft, lasted much longer. During the operation the membrane skin was taken from my mouth and grafted on to the eyelid edge. It made a lot of difference to my sight and was a lot more comfortable. The healing process took about four to six weeks. My mouth cleared up in no time and was never a problem. In fact, I complained more about my throat where they put the airway tube down my windpipe.

I am putting the third operation off until the Spring, as my surgeon has told me that an extended membrane graft around the corner of the eye might succeed much better, although it is possible that I will have to go through the second operation again.

Now all I can do is leave it to the hospital consultant, although I have been well cared for by all the medical team and it really does help when the hospital professionals know about my case with ED.
“Cool Hugs for Hot Little People”

We read with interest the following details in a ‘Cotton Comfort’ catalogue and thought they sounded brilliant for our little ones, whether they have eczema or not, who get hot at night, or in the car, or in the daytime, or in the pushchair ……..

The catalogue reads:

“Cool Puppy, Chilly Brown Bear & Arctic Sunshine. Soothe the hot itchy eczema with a snugly cotton friend who gives cool hugs for hot little people. Invaluable for bedtime. Each has a removable barley bag inside that can be cooled in the freezer to give around two hours of cool hugs. With rattle. Barley bag can also be microwaved to use as a hot water bottle. Each includes a zip lock freezer bag. Size approx. 30 cms,”

Made from: Cool Puppy 100% Cotton Fleece
Chilly Brown Bear 100% Cotton Towelling
Arctic Sunshine 100% Cotton Velour.

‘Cotton Comfort’ provides a range of pure cotton clothing, particularly for those with eczema. Ring 01524 730093, and ask for a catalogue.

Great for eczema – even better for our kids who don’t sweat.

They say Comedy is a great medicine!

Sarah Crosby asked her oldest friend, Nikki, if she would write an article for the newsletter giving her thoughts on what it’s like to have a friend with EEC.

Nikki writes:

Well, what can I say and write about Sarah!!!

I’ve known Sarah since age 11, at Ricky School. The one thing which stood out with Sarah, that I noticed, was her hair. That was the only thing I noticed; not her hands, toes or face, but her very blonde or white hair us girls would die for. Mind you, later on pink/multi-coloured hair, different colours for different weeks. Sarah was very confident and loved people’s attention, unlike myself.

When we left school we went around together a lot. I would get more annoyed with the stares that other people would give out, but Sarah told me to ignore it. I never understood why people stared, Sarah was just Sarah to me, no different to me or any others.

Black clothing; DM boots; festivals; pink hair – that was Sarah and still is, although the pink hair has gone (I think).

Sarah has lots of friends and still has loads of confidence, beautiful handwriting and very good at Art – no, she hasn’t paid me to say that.

When Sarah decided that she and Andy should marry, I sold my wedding dress to her, at low cost of course. It was lovely to see her walking down the aisle and I felt very proud. She looked lovely, it brought a lump to my throat.
Sarah has three lovely children and they arrived all very quickly – in more ways than one! Sarah didn't find out about the EEC syndrome until her second child was born. She has some of the symptoms relating to her condition.

We have always got on well and always have a great laugh talking about schools, what we got up to, drinking (and we still are) at pubs, going to fairgrounds, Sarah laughing at me being sick before a ride at the fair, spraying foam at me before my Art exam – and I failed ‘U’ grade – I blame Sarah!!

So, Sarah is Sarah to me – Confident – Loud (very) – Laughing – and A FRIEND.

A great friend to have – even if she has got EEC, she’s normal to me, and just Sarah.

Graphics Designer

Now that we are a charity, we need to reprint the leaflets to incorporate the charity number and the new website address. However, before reprinting we would like to ensure that the logo and layout of the leaflet are modern and up to date for the 21st century.

If you are a Graphics Art Designer or have skills in the field of Art and would be interested in helping us create a new logo, etc, would you please contact me in the EDS office to discuss this subject as soon as possible. Thank you.

Diana

Executive Committee Meeting 17th November 2001
Brief Summary of Minutes

Treasurer’s Report – Insurance for the charity to be researched. The Treasurer will draw up a framework for donations where the tax paid can be reclaimed through Gift Aid. A membership fee was agreed of £1 per person per annum, membership running from January to December. Membership guidelines will be drawn up.

Mission Statement – the following were agreed:
Strap line: “supporting a normal lifestyle”

Mission Statement:
“Our mission is to provide information, advice and support to those affected by Ectodermal Dysplasia”

Communication – Our website is now linked to the German and French ED sites, as well as the NFED. David Wyatt reported that there had been 778 hits over a 5 week period and he was proposing to run a Visitor Survey during December to establish more information on who was accessing the site, eg professionals, general public, etc.

Publicity – A Press Release is being produced. Publicity policy/strategy is being worked on to produce guidelines to help members when they are approaching, or being approached by, the media.

Update - Air conditioning has been obtained for a child who has ED and Cerebral Palsy, and Mandy White is working on another case at the moment.

Diana reported that 12 new members have joined EDS since the last meeting, 9 of whom are from UK, and 3 from USA. A geneticist, who has a particular interest in IP, has also joined the Medical Advisory Board.

Expenses – The Chairman will produce a policy for reclaiming reasonable travelling expenses, particularly in relation to the Medical Advisory Board.

Any Other Business – We are intending to have the ED leaflets updated and reprinted.
And finally …

2002 is going to be a very exciting and busy year for the Society as we begin to plan the best way forward. We will be

- creating a new database
- updating and circulating the leaflet
- circulating an article on ED to medical journals and newsletters
- updating our financial system
- etc.

All of this will only be possible with help from you and I am grateful to those who have already offered to help in different ways. However, I have a job for you all! As we embark on the challenge of being a charity and because we have got to have the leaflet reprinted to amend our details, I would really like you to consider the idea of having a more modern eye-catching look. Whether you think we should stay the same or whether we should move on, I would be really grateful for any thoughts and ideas you may have. In the article I have asked for the thoughts of a Graphics Art Designer, but I would really appreciate you all commenting on this.

We have many comments on how interesting and informative the newsletter is, but this can only happen with your input. Could you please put pen to paper and write about those wonderful experiences you have had as a family, as an individual, at school, at work, at your friend’s house, in the restaurant, in hospital, the happy times and the sad times……….these articles always speak to somebody, but can only do so if you write them down. Please let me have more articles.

Our Medical Advisory Board (MAB) continues to be a huge help and encouragement to the Society and I would like to thank them on your behalf for the time and expertise they give to us. We will be meeting with them again in March and will be putting together plans for the future of the medical side of the Society. In the meantime they are busy helping us to write and publish articles in medical journals informing the professionals in all different specialist fields of our existence.

You will see from the enclosed letter that the Conference has been postponed to the Easter holidays 2003. This is because I have had great difficulty in finding a venue with low cost accommodation to suit everyone. Hopefully you will all agree with my reasons, choice of dates and venue as explained in the letter. However, as set out in the Constitution, it is mandatory that the Charity hold an AGM every 12 months. As the conference will be nearly six months after the 12 month period for an AGM, we have received special permission to postpone our first AGM to coincide with the Conference. From April 2003 it is the Society’s intention to hold the AGM and Conference every year in the Easter holidays with the venue moving around the country.

I will continue to give a personal service, so whatever your questions small or large, please contact me and I will do everything I can to help. Please remember that as we grow in numbers and awareness, so do our resources and I am now able to seek and find more information and answers to your questions, but I can only do this if you ask!

Diana Perry

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