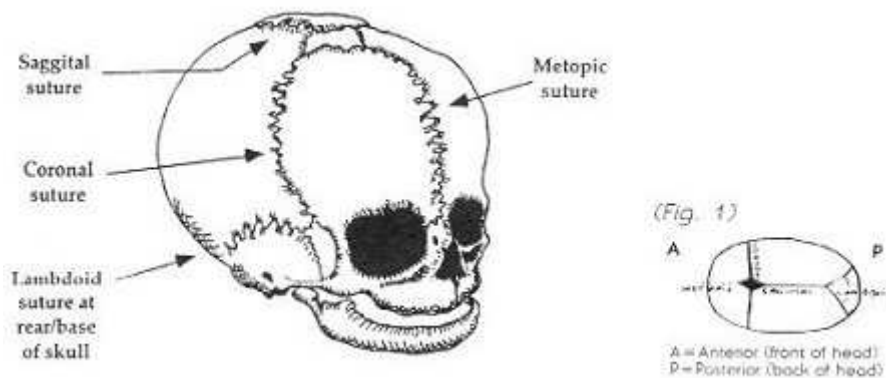


■ Non Syndromic Craniosynostosis (*Headlines factsheet*)

■ What is Craniosynostosis?

Craniosynostosis may be defined as the premature closure or fusion of the calvarial sutures occurring intra-uterine or shortly after birth. The calvarial sutures are lines of growth lying between the various bones of the skull. While there are a large number of sutures in the vault and base of the skull which can be involved this discussion will be limited to the common ones on the top of the head. These are six in number namely the **sagittal suture** which runs longitudinally down the midline of the skull between the anterior and posterior fontanels, the **metopic suture** which runs longitudinally from the anterior fontanel to the area between the eyes, the two **coronal sutures** - one on each side running transversely from the anterior fontanel to the area just behind the orbits and the two **lamboid sutures** which run obliquely downwards from the posterior fontanel to the areas behind the ears. (Fig 1)



The exact cause of sutures fusing prematurely in isolated instances is as yet unknown. The overall incidence averages out at approximately 1 in 3000 live births.

Under normal circumstances the growth of the individual skull bones occurs at right angles to the growing sutures. If a suture fuses prematurely the skull fails to grow at right angles to the involved suture(s). Importantly normal adjacent sutures respond to this growth restriction by increasing their activity and there is thus generally a compensatory growth in a direction parallel to the involved suture.

It is the failure of normal growth at right angles to the suture and the excessive compensatory growth at other sutures which gives rise to the classical skull shapes associated with the craniosynostoses. The typical shapes can be used clinically to predict the site of the abnormal suture.

These skull shapes have classically been given names which infer the involved suture and these will be covered at a little later in this discussion.

Because skull growth is most rapid during the first two years of life and continues to adulthood the presence of an abnormal or non functioning suture gives rise to a progressive deformity which is most rapidly progressive in infancy but has the potential to progress until growth is completed in adulthood. As mentioned there are sutures in the base of the skull as well which may be affected or, more frequently in the isolated synostoses, may grow compensatorially causing distortion of the lower orbits and the face at a later stage.

■ What other effects does fusion of a suture have?

In the vast majority of cases of single suture craniosynostosis the compensatory growth of the normal sutures is generally sufficient to allow the developing brain to grow without causing raised pressure. However, in a certain percentage of cases (somewhere between 10-15%) the restriction is such that the pressure within the skull rises (so called raised intracranial pressure) and this may cause functional problems in terms of development if left untreated. In addition there is evidence accumulating that there may be local

pressure effects underlying the involved sutures which may be corrected by surgery.

There are thus a number of reasons why surgery is generally indicated in craniosynostosis:-

- 1) For the treatment of an established deformity.
- 2) To attempt to prevent the significant progression of a developing deformity.
- 3) To relieve established raised intracranial pressure
- 4) To decrease the risk of developing raised intracranial pressure or other functional pressure related effects.

■ Types of single suture craniosynostosis

The type of surgery required is frequently determined by the degree of the deformity and the underlying sutures involved and therefore we will briefly review the different synostoses and their main features before proceeding to the discussion of surgery.

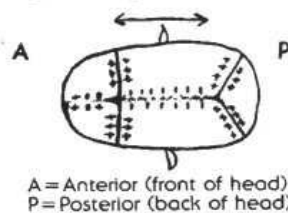
Sagittal synostosis (Fig 2) Fusion of the midline sagittal suture. Growth fails to occur across the head and excessive growth occurs from front to back. The resultant deformity is a narrow head which is excessively long. This is traditionally known by the term "scaphocephaly" which means "boat shaped head".

Metopic synostosis (Fig 3) Fusion of the anterior midline suture with failure of adequate transverse growth in the forehead and compensatory growth posteriorly and laterally. The end result is a central ridge of the forehead with a pinched look above the brows. The eyes tend to be fairly close together (hypotelorism).

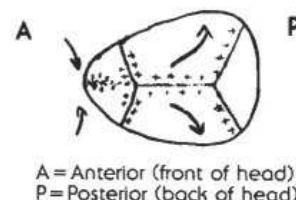
The overall skull shape (particularly anteriorly) is triangular in nature and this is classically known as "trigonocephaly" or "triangle skull".

Fig. 2 & 3

Sagittal synostosis



Metopic synostosis

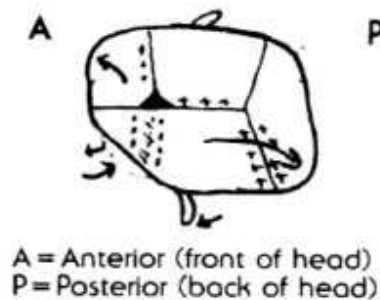


Coronal synostosis This may be divided into cases where only one coronal suture is involved or those where both coronal sutures are involved.

Unicoronal synostosis (Fig 4) is where one suture is involved and there is failure of adequate anterior posterior growth on the side of the involved suture. There is flattening of the brow and elevation of the upper part of the orbit and eyebrow area. The ear on the involved side tends to be pulled forward. The opposite forehead bulges significantly and there is increased growth in areas of the posterior skull. In addition there is

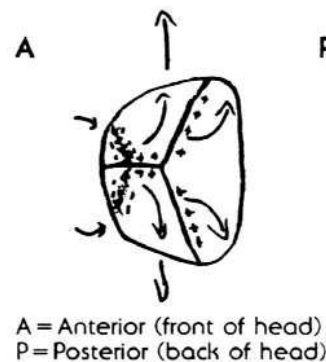
frequently compensatory growth in the cheek area of the involved side which in untreated cases may potentially cause facial distortion. The overall impression is of a skull which has been twisted skew, this condition is classically known as "plagiocephaly" or "oblique skull".

Fig. 4



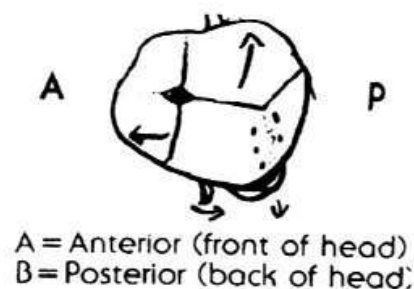
Bicoronal synostosis (Fig 5) is when both coronal sutures are involved. There is failure of adequate anterior, posterior growth on both sides of the skull. The skull becomes excessively wide and short from front to back. There may be excessive growth at the upper part of the forehead and both eyebrows and orbital rims are pulled up and are flattened. This short front to back appearance of the skull has given rise to the name "brachycephaly" or "short skull".

Fig. 5



Lamboid suture synostosis (Fig 6) This is rare with an incidence of approximately 1 in 10,000 live births. This presents with flattening of the skull around the involved suture with compensatory bulging occurring at the forehead on the involved side. Marked bulging of the mastoid bone behind the ear and a significant bulge on the parietal bone (which is the area above the ear) on the opposite side.

Fig. 6



■ Surgical Treatment

As already indicated there are a variety of reasons for preceding to surgery in craniosynostosis. Each case needs to be individually assessed in terms of functional indications and indications in terms of established or progressive deformity. Currently craniosynostosis surgery is one of the supra regionally designated services. There are four nominated centres:- The Radcliffe Infirmary, Oxford; Children's Hospital Birmingham; Great Ormond Street, London and Alder Hey Hospital, Liverpool.

Whilst the surgery required is fairly extensive, if performed in an established unit it is regarded as safe with acceptable risk in terms of the benefits achieved. These "benefits versus risks" are best discussed by the individual surgeons based on the individual features and indications in each child and it is not appropriate to give an analysis of all the various combinations in a general pamphlet of this nature.

In general as mentioned above the surgery is aimed at preventing progressive deformity, correcting established deformity and reducing any functional risk in terms of raised intracranial pressure.

It has been established that it is safe to remove segments of bone in the area of the involved sutures, to reshape segments of bone and change their position with predictable survival of the bone fragments and with reliable healing of the bone and soft tissue.

Sagittal suture synostosis is traditionally treated in the early phases by removal of some of the bone overlying the abnormal midline suture, the so called "strip craniectomy". This may be combined with various other ancillary procedures such as "plication" or tightening of the bones laterally to encourage the development of a broader, shorter head with growth. In established cases in older children more extensive reshaping of the skull may be required.

In the anterior synostoses (namely metopic, bicoronal and unicoronal synostosis) the aim is to recreate a symmetrical forehead and orbital rim. To "release" the area of the involved suture and there by allow more normal growth of the skull. The mainstay of this type of surgery is the "frontal orbital advancement and remodelling" procedure whereby the upper aspects of the orbits are freed and advanced unilaterally or bilaterally as appropriate and a more symmetrical forehead is reshaped from the existing or adjacent bone.

As mentioned lamboid sutures synostosis is uncommon and the surgery is aimed at preventing progressive deformity by releasing the suture and remodelling the posterior skull.

Irrelevant of the sutures involved if there is evidence of raised intracranial pressure then more extensive skull vault surgery may be required in order to expand the volume of the skull and thus relieve the pressure.

It is important to note, once again, that while this is major surgery it is generally classed as safe if performed by a multidisciplinary team ie a neurosurgeon with a plastic surgeon and/or a maxillofacial surgeon in an established unit.

Finally, it is important to note that because the underlying defect in the growth centre is as yet uncertain the surgery does not necessarily normalise the growth in all cases and there is a tendency in a certain percentage of cases for the condition to recur, or for progression to occur in other areas particularly the face, thus further operation or re-operations may be required in some cases. The predictive factors for this will depend on the sutures involved and the extent and rate of progression of the deformity. Once again it is impossible to give blanket guidelines for this and it should be discussed in detail with the treating surgeons.

***Written by Steven Wall. MBBCh(rand), FRCS, FRCPCH, FCS(SA) plast. ,Oxford Craniofacial Unit
Consultant Plastic and Reconstructive Surgeon, Radcliffe Infirmary Oxford, Feb 1997.***

Coping with Disfigurement (*Headlines factsheet*)

Introduction

"There are over 400,000 people in the U.K. with a scar; blemish or deformity that severely affects their ability to lead a normal life. " (Office of Population, Census and Surveys 1988).

The depressing statistics quoted above are not the words someone with a facial disfigurement or the parents of a facially-disfigured child wants to read. The last thing that they need to be told is that there are many people living disadvantaged lives in this country. However, such figures are undoubtedly useful in that they reveal a lack of adequate resources available to people with a disfigurement. They can also be used to argue the point that, if parents were given advice and help at an early stage then many of the difficulties they encounter could be prevented or at the very least overcome more easily. If specialist teams were on hand for parents to consult when and if they needed to, life could be made simpler.

Unfortunately such teams are only available on a limited basis and the "ideal" service does not exist (yet!) . There are, however, many individuals and groups, including people with facial disfigurement themselves, parents with disfigured children and professionals who are working together to make positive changes which could only have been imagined several years ago. There is now a real commitment to make sure that numbers quoted in statistics, like the ones above, begin to drop, although it must be accepted that this will not happen overnight.

Research has not only revealed areas which need to be changed, it has also shown that there are many people who are living with a disfigurement who are coping perfectly well and who deal with the demands life places upon them more than successfully. In lots of cases this can be through the use of self-help techniques and strategies which can be learnt individually, with friends, parents or as a member of a group. People can learn techniques and strategies which, combined with practical advice and information, they can develop and use on a day to day basis.

This leaflet is intended as a starting point. It does not offer a solution to all of your problems but it will give you some ideas and advice which you might find helpful. The information is intended for parents who have a child with a disfigurement and there is also a section for older children who are learning to cope with living with a disfigurement.

Coping with the attitudes and behaviour of other people

The experience of living with a disfigurement is not all doom and gloom but there are going to be times when you wished you had stayed at home, rather than deal with other people's ignorance! As the parent of a child with a disfigurement, feelings of stress, anxiety and at times loneliness and even despair may get on top of you. Our experience has shown that there are positive steps you can take to help re-gain control.

Firstly, you can gain knowledge about your child's condition. Talk to their consultant and don't be afraid to ask as many questions as you need to. Get them to write things down if you are worried about forgetting what they say. Go to the library and do some investigating yourself and keep an eye out for articles in newspapers and magazines.

Secondly, you can learn to use some simple strategies for dealing with difficult or embarrassing situations. These can also be used by your child and if they see that you are confident and happy to be with them they will develop their own sense of worth.

Although comments, questions and stares can be very wearing, especially when they come from total strangers, you will find that there are many people out there who are considerate and who will not judge your child by her/his looks. Hang onto this fact when you are dealing with the minority of people who are ignorant and rude.

You can choose your own techniques for coping with situations and you can adapt them to suit your own needs but here are a few ideas to get you started. Always remember that other people do not have the right to know about your child. Tell them what you want to - it's up to you.

What you say and what you do

Change the subject . . .

One ploy is to change the subject or distract their attention. If someone is staring at your child and you are unable to move away or you don't want to (e.g.in a queue at the supermarket or at a bus stop) you could try talking to them. Keep it simple - the weather may be a boring subject but it can be a useful one when you are trying to divert someone's attention from your child's face.

Explanations

You do not have a duty to educate anyone who asks questions about your child or make comments, but if you feel the situation would benefit from an explanation keep it brief and to the point. This kind of response can work well to prevent any further conversation. You could say:

"My daughter was born with a cleft lip, doesn't she look great!"

"My son has Aperts and is having some surgery which is helping him."

Speaking up for yourself. . .

You do not have to listen to comments which are upsetting for both you and your child, but if you want to say something in reply you can. Short responses are usually best (try to keep calm even if you don't feel it!) Here are some ideas:

"Don't stare, my child only has a scar on his face."

"My daughter has . . . you cannot catch it."

"Please don't stare, it makes me feel very uncomfortable."

Talk to your child and direct your thoughts away from the person/people who are commenting or staring. Use your own judgement in different situations, choose for yourself and your child the response you feel most comfortable with.

Positive affirmations

Make up short, positive sentences, together with your child if possible, and repeat them to yourself when you are in need of reassurance. Affirmations can say anything you want them to, they can be serious or silly, it's up to you. Some ideas could be along the lines of :

"I love my child because of who she is."

"Their words cannot hurt my child or me."

"Ugly is only a word."

It may seem strange to repeat sayings to yourself but it will get easier as you use them every day and they can be very useful to help build your confidence and self esteem. Practise your ideas with someone you feel comfortable with or stand in front of a mirror and test out new ideas.

Speaking clearly and holding yourself upright may seem simple techniques, but they can work extremely well when you are dealing with unpleasant or embarrassing encounters with other people. Don't worry if things do not work out first time. You will soon learn what is best for you.

Getting the support you need

There are going to be times when you need extra help. Don't be afraid to say "Yes please" when people offer to support you or even to ask for a hand when you're struggling. This will give you time for a break or the opportunity to do something you want to for yourself which is very important. Partners, family and friends are all sources of support. If you do not have anyone you feel you can rely on, ask your child's consultant or G.P. about support groups that may be operating in your area. You can share problems and worries in these groups with other parents who have first hand knowledge of your problems whether they be emotional, practical or financial. Parents in support groups may be able to provide some of the answers you are looking for and offer advice and assistance. Knowing you are not alone is an essential part of feeling in charge of your life once more.

Why do I look different?

Children begin to ask questions and become curious about their appearance from an early age. It is vital that you are open and honest with them regarding their disfigurement and any treatment they might need. This will help provide the information they need to cope with their disfigurement and to understand why some people react as they do. Use language that is easy to understand and experiment with drawings, stories and games to help you explain things. As they get older you can tell them in more detail about their condition. Use their own questions and your judgment to decide how much they can take in. Ask their opinion about treatment and surgery and whenever possible let them choose the option they want. Sometimes children want a break from surgery or they need to talk about their worries and fears. Listen to what they have to say and help them to work things through. There could be issues which they are trying to keep from you, especially if they are being bullied or teased.

Teasing . . .

Teasing is inappropriately defined in one dictionary as "tantalise" or "irritate", suggesting it is something that happens for fun. While this may be true between families and friends, it is also defined as "torment" which is much nearer to reality when used to describe the effect on a child (or adult) who has a disfigurement.

Take time to listen and talk to your child who may have anxieties and concerns about being teased or bullied. Ignoring it or pretending that it is not happening is not the answer for you or your child. When children are continually being picked on and reminded of their disfigurement and/or speech problems it can have a long term effect on their confidence and self-esteem.

You do not have to watch your child continuously but look out for changes in behaviour, emotional outbursts and uncharacteristic displays of aggression or withdrawal. Teasing is something that you and your child can work on together and you can help your child and ways to tackle it themselves. Short, easy to remember sentences that are effective could include:

"It's only a scar. "

"It cannot hurt you."

"I was born like this."

Teach your child that they do not have to tell people about themselves unless they want to. It's up to them what they say and with your help they will be able to work out the response that they feel happy saying. Remind them that, if people can see that they are confident and that they believe in themselves, others will realise that their words are not upsetting them and leave them alone

Older children and adolescents

This section has been written with the needs of older children in mind.

As you get older you are going to experience problems, both at school and in your everyday life. You need to develop the skills you have learned to deal with situations as they arise.

Bodies talk . . . !

Act and appear confident - even if you don't feel it. No one will be able to tell you are feeling nervous if you:

- Hold your head up. Don't look down at the ground - for one thing people can still see your face and for another it is dangerous. You might walk into something!
- Look at people. When you talk to someone, look them in the eye, be friendly and try to put them at their ease. You don't have to stare (you know how awful that feels) - look away now and then.

What you say . . .

It is important to stand up for yourself. This does not mean being rude and aggressive but if other people are saying things that are cruel and unkind you have the right to answer back. Some replies could be if you are feeling in a good mood and able to laugh at things):

- "thank you for telling me but I already know"
- "looked in the mirror yourself lately?"
- "tell me something I don't know"
- "at least people look at me, they'd never notice you"

You never know, you make someone laugh and they could become a friend.

Speak up, try not to mumble. If you have problems talking, just take your time and do your best. Stop and start again if you need to. Once you have the other person's attention they are more likely to listen to you.

If you cannot be bothered and don't want to talk about your disfigurement, change the subject or ignore the question altogether and the other person might realise that they should not have asked and talk about something else. If they go on asking questions you can be assertive and say :

- "I don't want to talk about that now"
- "I'm feeling tired and I'd rather talk about it another time".
- "let's talk about something else"
- "can we talk about you instead?"

Practise with a friend or a member of your family to see how it feels, what is OK and what makes you feel uncomfortable. Choose someone you can trust and who you know will support you. There are lots of replies you can make up yourself and it can be fun. Find ones which work for you. It may take time and a bit of effort. Keep them short and easy to remember. It will be worth it.

Remember, you do not have a talk about yourself unless you really want to. You can give people a long or a short answer or say nothing at all, it's up to you.

Bullying

When teasing stops and turns into bullying or bullying is the aim from day one, you must tell someone. Bullies pick on people who they see as being different and it is important that you do not let yourself become their victim. Use the skills which you have learned and always tell someone what is happening. They may think you will not tell anyone or even threaten you, but the only way to stop bullies is to get support.

There are many people who will be good friends and not judge you because of your looks - have nothing to do with the ones who treat you badly and make you feel down hearted and fed up.

This is only the beginning. You can learn to be in control and take responsibility for your life. Don't worry if something does not work first time. Your skills can be used again and again and the more you practise them, the easier it will get.

The end?

Of course not. As a parent you will be expected to deal with many different situations from birth, playschool/nursery, schooling and on to adulthood. You may want to wrap your child in cotton wool and protect them from the world, but that is neither possible or fair. Your child has talents like any other and has the ability and the right to make friends with other children and adults. There will be times when they need extra help and a shoulder to lean on and when you as their parent need help for yourself. This is par for the course and not a sign of failure or weakness.

With encouragement and support your child will learn to believe in them selves and develop their own personality - independent and responsible for their own lives.

Written by Julia Errington

Julia runs Face the Future; a counseling and support service for the facially disfigured and their families. Face the Future (reg. charity), c/o Skills for People, Key House, Jesmond, Newcastle-upon-Tyne NE2 3AT.
