a guide to understanding

crouzon syndrome

a publication of children's craniofacial association

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This parent's guide to Crouzon syndrome is designed to answer questions that are frequently asked by parents of a child with Crouzon syndrome. It is intended to provide a clearer understanding of the condition for patients, parents and others.

how can children's craniofacial association (cca) benefit my family?

CCA understands that when one family member has a craniofacial condition, each person in the family is affected. We provide programs and services designed to address these needs. A detailed list of CCA's programs and services may be found on our Web site at www.ccakids.com or call us at 800.535.3643.

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This booklet is intended for information purposes only. It is not a recommendation for treatment. Decisions for treatment should be based on mutual agreement with the craniofacial team. Possible complications should be discussed with the physician prior to and throughout treatment.

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what Is crouzon syndrome?

Crouzon syndrome is a deformity that occurs when some of the bones of the skull and face fuse or close abnormally. This abnormal fusion or early closure affects the skull and the upper jaw or maxilla. It may be inherited as a genetic trait or it may arise as a new condition in the family. In the familial type, the condition is inherited as an autosomal dominant condition. It will appear in each generation in the family so that one out of two children will have Crouzon syndrome. The severity of the condition varies from patient to patient.

how do I recognize this condition in my child?

The maxilla is retruded, which means that it fails to grow forward properly. Therefore, the maxilla, or upper jaw, and the upper teeth lie behind the lower jaw teeth. This situation is called a malocclusion. Although the upper teeth should be in front of the lower teeth, this condition causes the exact opposite positioning of the teeth. The medical term for this is a Class III (3) malocclusion.

The cheeks may be flat and the nose may be short. With a small maxilla, the orbits, or bony containers for the eyes, do not develop normally. In fact, they are too small front to back, which causes the eyes to be too prominent. This condition is known as exophthalmos. This produces prominent, staring eyes. The child may have trouble closing the eyes completely. There may also be eye irritation. Although the mandible, or lower jaw, grows normally and because the maxilla is retruded, it causes the lower jaw to appear enlarged or more prominent.

With some Crouzon patients, the areas over the top of the skull, from one side to the other, at the level of the ears, may also fuse and stop growing. This results in the slowing or halting of the forward growth of the forehead and the upper portion of the orbits. This further increases the severity of the deformity by flattening the front of the skull. Therefore, the eyes appear larger and more prominent. In addition, there may be increased difficulty in closing the eyes.

who is involved in the treatment?

In cases of a pure Crouzon syndrome, a pediatrician and a craniofacial surgeon should evaluate the child. An ear, nose and throat surgeon should see the child for routine ear examinations. It may also be necessary to consult a neurosurgeon and an ophthalmic surgeon. A social worker and craniofacial nurse coordinator also meet with the parents to discuss insurance issues and the need for additional help. They also explain aspects of clinical management, which may not be covered by clinicians. A dietician is available to give dietary advice and to answer questions about feeding, especially during the postoperative phase. At a later time, a pediatric dentist, an orthodontist, and an oral surgeon become involved in the management.

what treatment is available for crouzon syndrome?

After a general craniofacial examination, a treatment plan is established and other specialists begin certain examinations. Photographs are also made. Basic exams include the following:

- Dental impressions.
- X-rays including a panorex for the lower jaw position, cephalograms to assess the relationship of the upper and lower jaws, CT (computed tomography) scan to assess skull growth, orbital size and jaw relationships. These scans can be converted into vivid, three-dimensional images of the skull and facial bones.
- Hearing tests when possible since patients with Crouzon syndrome tend to have ear problems.
- An eye examination.

It is important for a geneticist to meet with the family to discuss whether the condition runs in the family. It is important for the child to be evaluated by a craniofacial team to provide support and treatment for patients and their families.

what other problems might we expect?

Some children will need ear drainage tubes inserted. In addition, if there is incomplete closure of lids causing the eyes to be severely exposed, a surgical closure may be performed by an ophthalmic surgeon. However, this is rare.

If CT scans show the skull is not growing fast enough for brain expansion and if the soft spot is closing, the head must be enlarged. The neurosurgeon and the craniofacial surgeon perform this operation. It is done under general anesthesia. An incision is made in the scalp in a zigzag fashion from behind one ear across the head to the other ear. This method allows the cut to be hidden in the hair.

During the surgery, the neurosurgeon removes the front part of the skull. The craniofacial surgeon removes the upper part of the orbits. Then the brain is able to expand. The two portions of the bone that are removed are then joined together with small plates. The plates will dissolve later.

The bony complex is put back in place but is advanced to increase the skull size to allow brain expansion. It is fixed in the new position again with plates which will dissolve. In some patients the front of the skull is of a strange shape and other cuts may be necessary to make this more normal. Then the scalp is closed and tubes are left in to drain the blood. If necessary, a bulky bandage is placed on the head and the child goes to the Intensive Care Unit. In most cases, normal nursing care is sufficient after a day or two, and the child will leave the hospital in three to four days. Frequently, no further skull surgery is required. If there are no indications for early correction, then the skull surgery can be done in combination with upper jaw surgery.

what else should I know?

When assessing the decision for upper jaw advancement, one must seriously consider the severity of the condition. If the maxilla is severely underdeveloped, then the face appears more sunken and the eyes seem more prominent. X-rays called cephalograms and panorex show the relative positions of the upper and lower jaws, as well as the arrangement and number of teeth. From this film and from dental impressions, the orthodontist will decide if there is a need for presurgical orthodontic treatment.

As a reminder, skull and jaw surgery may be needed. A scalp flap is fashioned and the upper jaw is freed from all of the surrounding soft tissue including the material in the eye sockets.

The cheekbones are exposed to the front of the ears. The neurosurgeon removes the front part of the skull. He then makes cuts on the roof of the orbits in front of the roof of the nose. This requires cutting around the orbital walls and through the cheekbones in front of the ears. The upper jaw is loosened at the back with a special instrument. Forceps are placed in the nose and mouth and then the upper jaw can be pulled into its correct position. Arch bars, which have been previously placed on the upper and lower teeth, allow the jaws to be wired together. Bone grafts are placed in the orbital gaps. In some cases, surgery may be performed on the lower jaw. This requires careful surgical planning. The front portion of the skull and supraorbital ridges are placed onto the upper part of the orbits and plated into position. Bony gaps can be closed with bone grafts from the skull. The scalp is closed and drainage tubes are placed to prevent accumulation of blood.

In most cases, the child is observed in the ICU for 24 to 48 hours. Then a few days of recuperation are necessary. Bruising and swelling are to be expected, causing the eyes to swell shut. The child receives nourishment intravenously, but will later be placed on a liquid or soft diet. As in all cases of facial deformity, additional surgical procedures may be needed on the bones of the face or the soft facial tissue.

what is meant by fixation?

The skull and facial bones are fixed, or stabilized, with plates. Although these are usually absorbable, metal plates are used in any area under severe stress. These can be removed during subsequent procedures. Occasionally, when indicated, wires may be placed.

what complications might be expected?

This syndrome requires major surgery and there can be major complications such as death or blindness. In the hands of an experienced craniofacial team, this is extremely rare. Collections of blood, infection, bony irregularities, and the necessity for further surgery can present problems. It should be remembered that these conditions are problems of growth and subsequent "re-correction" may be required.

where Is the best place to have my child treated?

Crouzon syndrome is a complex condition. It requires the expert skill of several different specialists working together. Craniofacial teams experienced in the management of these patients best treat these. Centers with craniofacial teams working together have the advantage of greater experience. This definitely leads to better results and fewer complications. In addition, ongoing research at these centers offers patients the latest break-throughs in treatment. As there are only a few experienced centers in the country, it is quite common for families to travel quite some distance to get the best care. Children who are treated locally by inexperienced teams or by individual physicians not working together as a team, are more likely to have unsatisfactory results. It sometimes requires two or three additional operations to correct what has been done. Another advantage of traveling to busy centers is the opportunity to meet other families and children affected with similar problems who can offer advice. These families often share their experiences, which provides moral support.

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