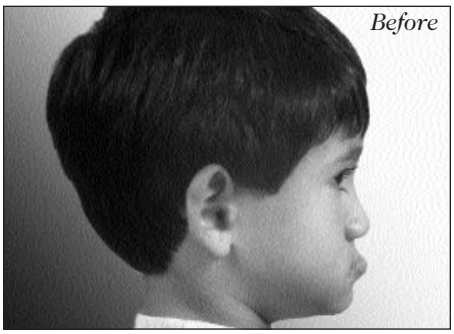
CCA News

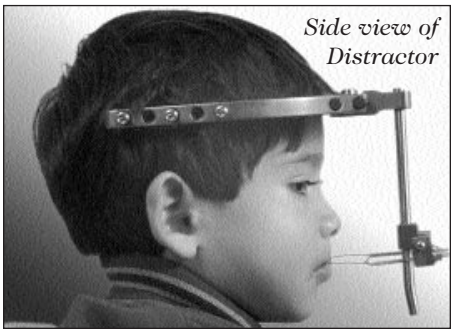
Newsletter of the Children's Craniofacial Association
CHER—Honorary Chairperson

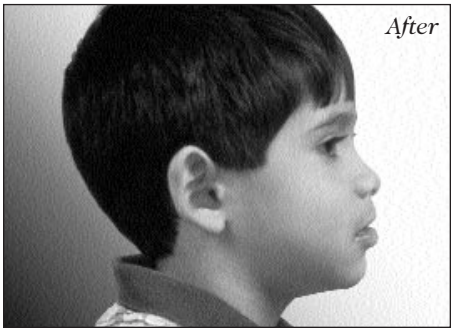
Winter 2000

What's New

Children born with syndromes such as Crouzon s and Apert s experience abnormal bone development in the areas of the upper jaw and mid-face. Until recently, lengthening the bones of the face required cutting and splitting the bone that needed to be lengthened. This is called an osteotomy. During the surgery, bone was harvested from another area of the body such as the ribs, hips, or skull. The harvested bone was placed in the gap. Titanium screws and plates kept the bone in place while the body healed and regenerated new bone.


Before


Side view of Distractor


After

Now there is an alternative bone-lengthening treatment. It is much less invasive and it provides more accurate results. This treatment is called Rigid External Distraction or RED. It is similar to the type of bone-lengthening technique used by orthopedic surgeons to lengthen leg bones. As was the case in the previous procedure, the craniofacial patient undergoes an osteotomy with small cuts being made in bone(s) that needs to be lengthened. However, instead of harvesting additional bone from another part of the body, a halo-like device is fixed to the patient s head and teeth with screws and wires. Each day the screws are tightened, which increases the gap where the bone was cut. Each time the gap is widened, the body naturally heals itself and compensates by regenerating bone to fill it. By turning the screws daily, within a six-week period the bone can be lengthened as much as 25 millimeters.

Aside from being less invasive, the RED system has other advantages over the traditional method of lengthening bone. It can be used to successfully treat patients at any age including childhood. Results are more accurate because adjustments can be made in many directions and at any time during the process. Best of all, patients report that this technique is painless.

The following are common questions regarding Rigid External Distraction:

Does the external distraction device hurt?

The rigid external distraction device (RED) is extremely well tolerated by the patient. This device is lightweight and

see **RED**, page 2

Message from the Chair

Happy Holidays! Our entire CCA organization wishes you and yours a wonderful, safe and rewarding holiday season. This has been an exciting and very positive year for CCA. We have been more successful in fundraising and have funds available for helping with travel expenses again. We are in the process of updating and adding to our syndrome booklets. Thanks to a tremendous amount of work by two of our families, the new Website is nearing completion. The board of directors had a strategic planning retreat and is working doggedly to execute it. Our premiere event for families, Cher s Family Retreat celebrated 10 years! 42 families attended this year and all had great fun. A Harley Davidson motorcycle was donated to CCA, raffled at the Sturgis rally, and provided a wonderful donation to our organization.

As you might expect, fundraising is always on the front burner. We ask you to make CCA your favorite charity, and to remember us when you consider giving. We now have a brokerage account set up to receive donations in stocks, which is handy for tax purposes. Please help us continue to serve those who need us and rely on us.

Terry Carmichael
Chairman of the Board

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there is virtually no discomfort experienced with the titanium fixation screws. It does not hurt the patient and no pain medications are required. This is the same experience reported for decades with the use of a cranial halo with neurosurgical and trauma patients.

Does the area with the screws need special care?

No. Patients should shampoo their hair and keep the scalp clean. Ointments, creams, or other medications should not be applied at the screw site.

Does the device limit the patient's activities?

Patients can engage in normal daily activities except for athletics, swimming, and contact sports.

Do the patients experience pain during the distraction procedure or when the screws are turned?

No, this process is pain free. The same is true with mandibular distraction.

Are the changes we see in these patients a true skeletal change or are they dental?

The changes are actually skeletal. With the use of the intraoral appliance as described, all of the vertical, horizontal, and transverse, or side to side, movements which occur during the distraction procedure are skeletal in nature. Rigid external distraction as described in this booklet produces no significant dental movements.

How much control do you have over skeletal changes in the upper jaw?

The RED allows for excellent horizontal and vertical control over the maxillary, or upper jaw, movements. The horizontal control is achieved through the activation of the distraction screws. The vertical control is obtained by changing the position of the horizontal cross-bar which supports the distraction screws. These adjustments can be performed any time during the distraction process.

How is the RED removed?

After the distraction process and the period of rigid retention, the titanium fixation screws are simply unscrewed and the RED is removed. This can be performed in older children and adults with local anesthesia in the clinic setting or with mild sedation in the younger age group.

Is any special care required at the site of the titanium fixation screws following removal of the device?

No. Normal scalp hygiene is continued and the small separation of the skin at this site closes within 24 to 48 hours.

Can the patient eat and speak with the rigid external distraction (RED) device in place?

Yes. The day after surgery patients are placed on a soft diet and they gradually progress to a harder diet. Swallowing and speech are not impaired.

The manufacturer of RED, KLS-Martin provided information for this article. This article is not an endorsement and intended only to inform.

Share the Magic!

Send flowers for the holidays!

Call **1-800-OUR GIFT** (687-4438) or visit **www.OurGift.com**. Every time you order flowers through Our Gift, 10% of your purchase is donated to CCA.

Meet 11-year-olds Tony and Chris Sapp

Chris (left) and Tony (right) Sapp

Q. Where were you born?
A. Palm Beach Gardens, FL

Q. Where do you live now?
A. West Palm Beach, FL

Q. What grade are you in school?
A. Sixth grade

Q. What are your hobbies?
A. We like to play Nintendo, watch auto racing and football, and collect Nascar memorabilia. Chris also likes to play the keyboard.

Q. What other activities do you enjoy?
A. We both enjoy playing baseball and soccer and helping our family and friends. We also like to help take care of our niece, Mena, and nephew, Austin.

Q. What do you want to be when you grow up?
A. We want to own our own business, maybe a bed and breakfast.

Q. What kind of craniofacial condition do you have?
A. We both have Apert Syndrome. We used to be the only identical twins we knew of with this. But now we know there are twin girls in Kansas who also have Apert Syndrome.

Q. How many family retreats have you attended?
A. We have been to four family retreats.

Q. Which was your favorite?
A. They were all fun, but Denver, Colorado was the greatest. Cher was there and she is always so nice to everyone. We also got to meet Reggie Jackson.

Q. What has growing up with a craniofacial condition taught you?
A. We might look a little bit different, but we like to do the same things that other people do. People should treat us just like they would want to be treated. Some people might stare at us, but they shouldn t. They could ask us questions and smile instead.

Attention Deficit Disorders: What are they?

Kristen E. Bolomey, Psy.D.

During the past several years, attention deficit disorders have received a great amount of press and seem to be diagnosed more and more often. Is this because it has become a "fad" disorder or because the characteristics are better understood? The answer is "both."

Attention deficit disorders include the diagnoses of: 1.) Attention-Deficit/Hyperactivity Disorder — Predominantly Inattentive Type, 2.) Attention-Deficit/Hyperactivity Disorder — Predominantly Hyperactive-Impulsive Type, 3.) Attention-Deficit/Hyperactivity Disorder — Combined Type, and 4.) Attention-Deficit/Hyperactivity Disorder Not Otherwise Specified.[1] Difficulty paying attention can include problems in starting activities, maintaining concentration, resisting distractions, trouble with organization and motivation, and difficulty with transitions from one concept or activity to another.[2] Difficulty with impulsivity involves problems thinking through consequences before acting, impatience, poor tact, poor anger control and poor frustration tolerance.[2] Whether an individual experiences one or both of these characteristics, his or her academic, social, occupational, and emotional functioning will be affected.[3]

Attention deficit disorders are organically based disorders that are often inherited. These disorders involve neurotransmitters and/or the structure of the brain. Attention deficit disorders occur before the age of seven years, and may be congenital (the child is born with it) or the result of a brain injury.[1] This type of injury may be the result of birth trauma, maternal substance abuse during pregnancy, traumatic head injury, or exposure to toxins. One reason for the increase in the incidence of attention deficit disorders over the past five to ten years is the increase in the number of premature infants who have been able to survive, as well as the number of babies born with congenital abnormalities that can be successfully treated. Children who are born at 24 or 25 weeks gestation (full-term is 38-40 weeks) are surviving at rates that would have been unheard of five years ago. Although these children have survived infancy, there may be a price to being born with an immature nervous system.

Children born with craniofacial abnormalities are particularly at risk given the atypical growth of the head, which can affect brain development. Some disorders include visual or hearing deficits, which make paying attention even more difficult, however, attentional difficulties that are primarily the result of hearing or vision problems are not considered attention deficit disorders. Numerous surgeries can also cause delays in development, although these are thought to be temporary in nature.

Unfortunately, attention deficit disorders are not always straightforward diagnoses. Children who are experiencing depression, anxiety, or who have recently been physically or emotionally traumatized may exhibit symptoms quite similar to an attention deficit disorder.[4] It is very important that these disorders be diagnosed appropriately because the treatments for each are very different. Children have not yet developed the coping skills that adults have. While an adult may be able to talk to someone about a problem, ask for help, use physical or creative energy to deal with frustration or attempt to work toward problem solving, a child may only recognize that he or she is uncomfortable. This may then lead to acting out behaviors.[5] Because attention deficit disorders are so popular now, many times stimulant medication is prescribed before an adequate history or assessment can be made. If a child is really experiencing depression, anxiety, or even hyper- or hypothyroidism, this approach may delay or even mask the problem. Ruling out hearing and visual problems is absolutely essential as is a psychological evaluation.[3] A psychological evaluation completed by a psychologist (usually someone with a Ph.D., Psy.D., Ed.D., or an M.A./M.S. in psychology) can determine whether an individual s attentional problems are the result of emotional issues, a learning disability or other neurological factors. This type of assessment can also aid in educational planning. Psychologists do not prescribe medications so a psychiatrist or neurologist would have to be consulted if stimulant, anti-depressant, or anxiolytic medication were to be considered. For children under 14, it is necessary to involve pediatric specialists.

Children with craniofacial issues who also have an attention deficit disorder may also be experiencing difficulties with self-esteem and self-confidence associated with disfigurement. Although this does not usually occur until middle childhood (about 8-10 years), it can contribute to sadness and anxiety that can increase attentional problems. Counseling or psychotherapy may help a child work through frustrations while learning to organize him or herself.

Children with attention deficit disorders that are not treated are more likely to drop out of school, have more grade retentions, may be more likely to use drugs or alcohol and have lower academic skills.[2] However, there is hope. With appropriate assessment, medication and intervention, children can learn to develop behavioral controls that were difficult for them to achieve. Medication is likely to help control behavior that the brain was not capable of managing on its own. Then when behaviors are maintained, therapy can be beneficial in teaching coping skills. Attention deficit disorders are not diseases that are cured, but rather disorders that must be approached strategically in order to maximize the individual s, and his or her family s quality of life. This is not the result of poor parenting or willful behavior on the part of the child. By realizing this and working together, the child with the attention deficit disorder, his or her family, teachers and friends will enjoy a greater sense of well being.

References:

[1] Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (1994). American Psychiatric Association: Washington, D.C.

[2] Robin, Arthur L. (1995 presentation). Surviving ADHD in Adolescence: A guide for practitioners on assessing, diagnosing, and treating the adolescent with attention deficit hyperactivity disorder.

[3] Barclay, Russell (1998). Attention-Deficit Hyperactivity Disorder: A handbook for diagnosis and treatment. Guilford Press: New York.

[4] Hallow, Edward and Ratey, John (1995). Driven to Distraction: Recognizing and coping with attention deficit disorder from childhood through adulthood. Simon and Schuster: New York.

[5] Lott, Lynn; and Intner, Riki (1995). The Family that Works Together Turning family chores from drudgery to fun. Prima Publishing: Rocklin, California.

3 Cheers for Volunteers

Did you know that Children s Craniofacial Association has a gift shop? It is located in the North Park Shopping Center in Dallas, Texas and is called **Our Children's Store**. The store is a collaborative effort benefiting 50 area children s charities and it is run by volunteers from each of these charities. Our Children s Store is chocked full of a variety of unique gifts suitable for any occasion and for people of all ages.

One day each month, CCA volunteers man the store. **Cathy Evans**, CCA s Volunteer Chairperson, is responsible for recruiting and scheduling our workers. Her commitment to CCA is a very personal one since her daughter was born with a cleft lip and palate. The store is just one of many ways that Cathy serves CCA and its families. Cathy works as Office Manager for Dorfman Productions, an oil production company in Dallas.

Others volunteering their time and energies to Our Children s Store include Cathy s daughter, **Lisa Evans**, who has served as a CCA volunteer for many years, **Suzanne Nordmann**, a freelance artist, **Susan Cooper**, a community volunteer, and **Bruce** and **Robin Williamson**. Bruce is a digital illustrator for Rapp Collins Worldwide and Robin is a graphic designer and serves on CCA s Board of Directors.

These folks are the foundation of our volunteer corps. We take off our hats to these dedicated volunteers!

Call Cathy Evans at work at 214-361-1660, at home at 972-636-9497 or call Our Children s Store at 972-240-7730 for information regarding volunteering.

Check Out Our New Website!

We have a new look and address for our Website: **www.CCAkids.com** (the old address will be rerouted to the new). There will be a lot more news and information available on the new site.

Please go on our site and register. If you d like to receive future newsletters via Internet download (a .pdf file), please indicate this on your registration. It will save CCA money in printing and postage to deliver information to you in this format.

Many thanks to **Brenda Malloy** and the crew at **MasterLink** for handling our Website in the past.

Thanks to our hard-working committee that got this site up and running: **Fred** and **Rose Seitz**, **George Beavers,** all of **Directed Technologies, Inc.** (They donated all their time and talents in the construction of the Website.); **Mark** and **Donna Shea** (Mark and Donna have been active in our Website in the past and will continue to act in the capacity of information point people on the new Website. Any info or questions about the site, please contact them.); and board members **Robin Williamson** (Robin designed the new look for the site and acted as committee chair.) and **Richard Wallets**.

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Cher

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