CROUZON'S SYNDROME: A DIFFERENTIAL DIAGNOSIS

by

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June 1990

Presented to The Oregon Health Sciences University School of Dentistry, Department of Pediatric Dentistry in partial fulfillment for a certificate in pediatric dentistry

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ABSTRACT

Crouzon's syndrome is craniosynostotic disorder which is inherited in an autosomal dominant pattern. There is often severe facial deformity associated with this syndrome as well as other problems. In this paper a literature review is presented along with a case report of an affected child.

The purpose of this paper is to distinguish Crouzon's syndrome from the other syndromes that share craniosynostosis as a significant finding. Many features of Crouzon's syndrome will be examined in depth. This will include pathology of the various organ systems, genetics, psychological profiles and problems as well as a look at the historical and modern techniques of surgery. As there are over sixty separate syndromes that feature craniosynostosis, I have chosen two, Apert syndrome and Saethre-Chotzen syndrome, to compare and contrast selected elements with those of Crouzon's syndrome. The reasoning behind this is that these two syndromes are frequently confused with Crouzon's syndrome. The paper will then be completed by a case report of a twelve year old, Darryl Fortier, who has Crouzon's syndrome which was surgically corrected at a young age. This case report will center around the characteristics examined in the literature review.

LITERATURE REVIEW

It was in 1912 that Dr. O. Crouzon first presented his now famous study, Dysostose cranio-faciale hereditaire, to the Medical Society of the Hospitals of Paris. His report was based on two patients, a twenty year-old mother and her three year old son. These two subjects both showed manifestations that would later be termed "Maladie de Crouzon" or Crouzon's syndrome.

The characteristic findings of this syndrome are brought about by a cranial synostotic malformation. These findings typically include bilateral exophthalmus with external strabismus, psittichorhina, which is a parrot beaked nose, and mandibular prognathism. Crouzon went on to point out that this syndrome followed a familial occurrence.

In the years since Crouzon's original report, many more cases of this interesting syndrome have been noted. Other findings, such as maxillary hypoplasia and a low hairline (Juberg, 1973), have been added to the diagnosis of Crouzon's syndrome. Other forms of craniofacial dysostosis have also been described with subtle features separating them from true Crouzon's syndrome. One example of this is Apert syndrome. Like Crouzon's, Apert syndrome or acrocephalosyndactyly, results from a defect in the cranial sutures. In addition to the main features noted in Crouzon's syndrome, patients with Apert syndrome exhibit syndactyly of the hands and of the feet, often combined with anomalies of various other organ systems. Apert syndrome is closely related to Crouzon's syndrome in the sutures affected and the degree of craniosynostosis involved. Like Crouzon's, craniosynostosis is rarely absent. It is the rarest of the three syndromes under consideration in this paper with an estimated frequency of 1:1000,000 to 1:2,000,000 versus Crouzon's 1:25,000 (Caronni, 1985; Cohen, 1986).

Saethre-Chotzen is another syndrome whose most significant finding is craniosynostosis. It is relatively common among this group of syndromes. Saethre-Chotzen syndrome so closely resembles Crouzon's syndrome, It is often difficult to differentiate between the two. Patients with Saethre-Chotzen syndrome can range in severity from presenting with almost no clinical abnormalities to having an appearance that mimics severe Crouzon's.

In addition to craniosynostosis, Saethre-Chotzen patients share a frequent low set frontal hairline, hypertelorism and strabismus with those patients afflicted with Crouzon's syndrome. Individuals with Saethre-Chotzen syndrome can also present with maxillary hypoplasia and a relative mandibular prognathism. Unlike Crouzon's syndrome, where the incidence of synostosis is almost one hundred percent (Dodge, 1958), patients affected with Saethre-Chotzen syndrome may or may not show this anomaly. When present, craniosynostosis is quite variable in onset and degree. Saethre-Chotzen patients also frequently show asymmetric synostosis, producing plagiocephaly and facial asymmetry. These patients often have abnormalities of the hands and of the feet similar to Apert syndrome. These abnormalities are generally not severe, often manifesting only as a partial cutaneous syndactyly, usually of the second and third fingers.

The pathology noted in Crouzon's syndrome is, as mentioned before, the result of a defect in the cranial sutures. This defect causes early synostosis of all or some of the sutures. The sutures most likely to close first being the coronal, lambdoidal and the sagittal. This results in growth of the skull in a direction in which the sutures have yet to close. The unplanned growth noted above gives the skull a shape that's best described as being a combination of scaphocephalic (boat shaped) and trigonocephalic (wedged shape).

The skull in Crouzon's syndrome is widest in the temporal regions and somewhat flattened in the parietal regions. Actual head circumference is decreased. The sella turcica is enlarged when viewed on lateral headplates.

The skull in patients suffering from Apert syndrome has an accentuation of the digital markings which is also found in Crouzon's syndrome. Hyperacrobrachycephaly,which is a skull characterized by being tall and flat, is common. The forehead is somewhat steeper then is noted in Crouzon's with a horizontal groove being present in childhood. The cranial base is malformed and may have a significant degree of asymmetry.

The orbits in patients with Crouzon's syndrome exhibit much pathology in their formation. The orbital roof is shortened while the height of the orbital opening is increased. Both the lateral and inferior orbital margins

are quite retruded. Overall this gives the orbits a greatly reduced capacity, hence the exophthalmus.

The orbits in Saethre-Chotzen syndrome tend to have a near normal capacity while those found in Apert syndrome fall between the other two syndromes, exhibiting moderately reduced capacity (Caronni, 1985)

The mandible in Crouzon's syndrome has a narrow ramus and is shorter overall when compared to normals (Kolar, J.C. 1988). It also tends to be backward rotated with a steep mandibular plane. The reason these patients appear to be prognathic is not the shorter mandible, but the presence of a hypoplastic maxilla. This maxilla is generally small and also in a retrognathic position on the cranial base. Extensive maxillary remodelling occurs but, does not compensate for the synostosis of the maxillary sutures.

Due to the altered relationship of the maxilla and the mandible, severe malocclusions are the rule with these individuals. All patients tend towards a class three dental relationship with mandibular prognathism. This disharmony tends to worsen as the child enters puberty. Bilateral or unilateral (most often with a shift) crossbites are often present (66%) (Cohen, 1986). The hypoplastic maxilla gives a shortened maxillary dental arch. Arch width is also somewhat reduced. This is partially due to the mid-palatal and transverse palatal sutures undergoing partial to complete

fusion. This restricted arch form along with frequently present (50%) lateral palatal swellings give the appearance of a highly arched palate. When actually measured the palatal height did not differ significantly from normal (Cohen, 1986). This is in contrast to Apert syndrome where narrow high arched palates are often found.

Severe crowding of the maxilla is common. Crowding seems to be most severe in the premolar area. Ectopic maxillary first molar eruption occurs in a full fifty percent of the patients (Caronni, 1986).

Apert syndrome has oral findings that are often more severe then those found in Crouzon's. While both syndromes exhibit class three malocclusion, that found in Apert syndrome is worse, often with an anterior open bite in addition to severe crowding. Oral anomalies found in Saethre-Chotzen syndrome include highly arched palate, cleft palate, malocclusion, supernumery teeth as well as defects of the teeth themselves (Bartsocas, 1970; Gorlin, 1964).

Crouzon noted several anomalies of the teeth. These included aplasia of single teeth, abnormal premolar morphology, and shovel-shaped maxillary incisors. These problems were later found to exist with the same frequency as found in the general population, and thus is not a significant finding in Crouzon's syndrome.

An airway that is somewhat restricted is a common abnormality in

Crouzon's syndrome. The nasopharynx is reduced in all dimensions requiring the patients to carry their heads in an extended position to protect the airway. This partial nasopharyngeal obstruction also causes many patients to be obligatory mouth breathers. In one study 32% of the subjects exhibited obligatory mouth breathing (Cohen,1986). In severe cases of the syndrome a combination of basilar kyphosis and arrested maxillary growth can bring about occlusion of the epipharynx resulting in respiratory distress. Congenital tracheal stenosis is another cause of respiratory distress in Crouzon's syndrome.

The eyes of patients suffering from Craniofacial Dysostosis exhibit exophthalmos and hypertelorism due to the abnormal skull growth and from shallow orbits. The orbital region tends to show the greatest amount of dysmorphology in the entire craniofacial complex. The intercanthal, eye fissure and biocular lengths are all large when compared to normal values. Originally it was thought that the exophthalmus was due to an increase in intracranial pressure but, this proved not to be the case and most authors now agree that the cause is the rather shallow orbits.

Exophthalmos or proptosis is a constant feature in Crouzon's syndrome. The suggested causes of proptosis are many and include the following: (1) A reduced length of the orbital floor secondary to arrested maxillary

growth. (2) Displacement of the greater wings of the sphenoid bone in the anterior direction. (3) A short anterior cranial base. (4) An increase in the volume of ethmoidal air cells (Hemmer, 1987). The extent of the proptosis can be so severe that exposure conjunctivitis and keratitis become apparent. There have been multiple cases of actual luxation of the eyeglobes.

Exotropia (divergent strabismus) occurs in approximately 80% of the cases (Greaves, B., (1979). This has been attributed to the shallow orbits as well as to other bony pathology. This exotropia is of a very characteristic type in Crouzon's syndrome with the eyes showing exotropia while the eyes are upturned and a normal gaze when downturned. When viewed from the front these abnormal eye movements present a "V" pattern.

One half of patients with Crouzon's will present with visual loss (Cohen M. 1986). This may exist at the time of birth, develop slowly, or progress rapidly-occasionally resulting in complete blindness. Visual loss can be due to the increased intracranial pressure noted in these patients, or to constriction of the optic canals by bony pathology.

Exposure keratitis noted above is often responsible for severe visual impairment. Infection and scarring of the cornea can occur leading to decreased corneal sensation. This leads to further trauma and eventual

loss of visual acuity.

The evidence for a genetic role in Crouzon's syndrome is overwhelming. To fit Crouzon's original criteria the syndrome must follow a hereditary occurrence. Approximately 70% of cases in one study were found to be familial with the remaining 30% being sporadic in nature (Atkinson, F.R.B. 1937). Other studies have found the reverse with the majority of cases being sporadic (Kreiborg, D 1982). One possible explanation for the relatively small number of inherited cases is that people suffering with this syndrome tend not to marry and reproduce due to the severity of the facial defects present.

Crouzon's syndrome occurs in white, black, oriental as well as other populations. The actual frequency is difficult to determine due to the clinical appearance being similar to other syndromes whose main finding is craniosynostosis. The best estimate available is 1 in 25,000 of the general population. This number is based on the studies of several authors (Cohen,1986; Shiller,1959).

The inheritance of Crouzon's syndrome is autosomal dominant. There have been several reports of autosomal recessive cases of this syndrome but, these have largely been proven to be either other syndromes or autosomal dominant in inheritance. One study looked at an Amish population that had a large number of affected individuals (Cross and

Opitz 1969). The inheritance in this case appeared to be autosomal recessive. This same population was examined seven years later and the affected individuals were found to have Jackson-Weiss syndrome, an autosomal dominant condition that can appear quite similar to Crouzon's syndrome (Jackson and Weiss 1976).

Variability of expression is another problem when the inheritance of Crouzon's syndrome is examined. A family with several members showing different degrees of Crouzon's syndrome were examined. One child was severely affected, showing prominent proptosis and the kleeblattschadel or cloverleaf skull anomaly. The child's siblings were either not affected or showed only moderate manifestations of the syndrome. When the mother was examined it was found she did have the syndrome evidenced by ocular proptosis but, she had no recognizable craniosynostosis (Shiller 1959).

Both Apert syndrome and Saethre-Chotzen syndrome also follow autosomal dominant modes of transmission. This, at times, has been difficult to establish in Saethre-Chotzen syndrome as variable expressivity and incomplete penetrance are common.

The psychological impact on parents and the patient affected with Crouzon's syndrome can be both enormous and devastating. The birth of the child is often a major crisis, disrupting parent-infant bonding.

Parents of malformed infants tend to pass through an identifiable sequence of emotional reactions beginning at the child's birth (Arndt, 1986). The initial stage is one of great shock. This stage quickly passes into the second stage of disbelief. Parents tend towards denial in this stage. The third stage is marked by feelings of anger and sadness. This stage can last a long period of time but, gradually passes into the final stage of acceptance. The parents are now more comfortable with their child's syndrome and it's associated problems. Counseling by an individual familiar with the situation has been shown to be of benefit. It is important that the parents avoid rejecting the child as well as not becoming overprotective. Other problems that have been reported and whose incidence may be decreased by professional guidance include chronic sorrow, social withdrawal, alchohlism, psychosis, divorce and child abuse (Arndt 1986).

As the children become aware of their malformations they develop different coping methods to protect their self-esteem. They can try to ignore their defects and center on the positive attributes they posses, or dismiss those who tease them as cruel or shallow (O'Donndell 1985). Affected children oftentimes can be cheerful and outgoing until they reach school age. At this time they are usually subjected to an increasing amount of teasing and ridicule, forcing the child to become more self

conscious of his or her appearance. This oftentimes leads to a child who is withdrawn and severely lacking in self-confidence (O'Donndell 1985). As the children approach their early teen years their self-image is further degraded. Rejection by peer groups become the norm. At this point open depression usually becomes apparent (Landers 1989).

It is entirely possible for mildly affected Crouzon's and Saethre-Chotzen patients to avoid much of the psychological suffering noted above. This is usually not the case with Apert syndrome due to it's severity. In addition to having more extensive facial deformities, Apert patients exhibit fusion of the fingers and of the toes. This further alienates these people as they can have difficulty with the simplest of manuel tasks (Caronni, 1985).

Menal retardation is yet another problem that Apert syndrome patients must deal with. While both Crouzon and Saethre-Chotzen patients can exhibit retardation, it is much more prevalent in Apert syndrome. Approximately one half of individuals with Apert syndrome will have normal to near normal intelligence with the remainder showing a marked reduction in IQ (Patton,1988).

Surgical correction of facial deformities can give these patients a much improved self-image (Palkes 1986). The timing of surgeries is determined by the child's emotional state, social awareness, surgeon's assessment and oftentimes parental pressure (Ousterhout 1986). The

general feeling is the earlier the better to prevent many permanent psychological scars.

Surgical correction of the craniofacial deformities associated with Crouzon's syndrome serves two important functions. First, extensive surgery is oftentimes indicated to correct cosmetic deformity. Second, it is believed that "unlocking the brain" will lessen the chance of mental deficiency occurring (Lane 1892).

The concept that early surgery can prevent brain damage has much controversy over the years. Several studies have noted increased intracranial pressures associated with fused sutures. In one of these studies seventy-five children were evaluated using an epidural pressure transducer for at least twelve hours. The children were all pre-surgery, age six to fifteen years with various forms of craniostenosis involving one or more sutures. It was found that the more fused sutures noted, the higher the intracranial pressure (Renier, 1982). The younger children tended to have higher pressures, likely due to the effect of brain growth inside a non-expanding skull. As the children became older and brain growth slowed, these higher pressures became normal (Cohen 1986).

Other studies have noted no post surgical changes in mental status. Today the majority of specialists in this area feel that a single fused suture may cause an increase in intra-cranial pressure but, the likelihood

of this causing a mental deficit is small. However, as the number of fused sutures increases the probability of a deficit increases (Renier 1982). Craniostenosis does have a great effect on the shape of the brain. This has been thought to be a cause of retardation but, has been difficult to prove.

The first surgery to correct cranial stenosis was performed on a microcephalic child in 1888 (Lane 1892). The operation consisted of the excision of a single suture that was believed to be the cause of the patients condition. Although the patient died within twenty four hours, the surgery sparked much interest in the medical community. Several successful craniectomies were said to result in an improvement in the patients mental status. The majority of these early surgeries centered around the excision of the fused suture.

Several techniques were developed that aided in preventing the recurring problem of the excised sutures from growing back together. This was believed to allow the brain full development as it was no longer enclosed in an immovable chamber. One of these techniques involved the application of Zenkers solution, a cauterizing agent, directly to the dura. It was believed that this would destroy the bone forming elements present. Although successful when just suture patency was examined, the severe side effects eventually led to its demise. Other techniques

featured the implantation of tantalum foil or polyethylene film to combat the problem of premature closure of excised sutures (Simmons 1947).

In the 1960's, Dr. Paul Tessier introduced new surgical methods to correct different types of craniosynostosis and their associated cosmetic defects. His methods included radical reconstruction, often of the entire face, while at the same time opening the fused sutures. Using methods such as these present day surgeons can obtain normal configurations in greater then ninety percent of the worst malformations (Tessier 1971).

CASE REPORT

Darryl Fortier is the product of a thirty nine week uncomplicated pregnancy. He was delivered by C-section for failure to descend after administration of pitocin. Polyhydramnios and heavy meconium staining were noted. His birth weight was 4022 gms. (75%-90%), length 54.6 cm. (90%-97%), and head circumference 36 cm. (50%). The only neonatal problem noted was mild tachypnea, likely due to the heavy meconium noted. The maternal age was twenty four years with the paternal age being twenty eight years. The weight gain with this pregnancy was sixty five pounds of which forty pounds were lost by two days post delivery. During the first trimester there was exposure to amphetamines. The mother smoked greater then twenty cigarettes per day throughout the pregnancy. There was a history of occasional alcohol use.

At birth Darryl was noted to have an unusual cranial contour with mild oxycephaly and prominence of the forehead. Coronal suture synostosis was suspected at this point and was substantiated by skull radiographs. The diagnosis of Crouzon's syndrome was made.

Upon this diagnosis other members of the Fortier family were examined directly and indirectly (via pictures). It was found that evidence of craniosynostosis went back as far as four generations. Darryl's father as well as his aunts, grandmother, great grandfather, and great great grandmother all had features of a craniosynostotic syndrome. Other features noted in affected family members included orbital hypoplasia, proptosis, and brachydactyly.

At age eleven months the decision was made to surgically correct Darryl's craniosynostosis. This surgery consisted of opening up the coronal sutures. The surgery was performed at The University of California, San Francisco Medical Center.

Darryl's next full physical exam was performed three months later at the UCSF Genetics Clinic. The following list includes the findings of this exam.

Age - 14 months.
Height - 31 inches. (50%)
Weight - 29 pounds. (97%)
Head circumference - 51 cm. (98%)
Cranium - Frontal prominence, prominent bitemporal area, mytopic

sutures open, surgical suture line (coronal), shallow orbits.
Forehead - Prominent glabella, prominent forehead.
Ears - Length (R&L) - (50%), poorly formed helices, overfolded helices (R&L), lateral helices flattened (R&L).
Eyes - Inner canthal distance - 2.8 cm. (75%-90%), interpupillary - 5.5 cm. (98%), outer orbital - 9.5 cm., bilaterally epicanthal folds, no detectable visual problem.
Nose - Flat bridge.
Face - Slight hypoplasia of malar region.
Midface - Mildly small mandible.
Mouth - small.
Oral cavity - High palate.
Hands - Palm length - 5.5 cm. (50%), third finger length - 3.5 cm. (10%), brachydactyly (mild), fingers tapered.
Feet - Toes short, clinodactyly of toes.
Neurological - Reflexes normal.

Darryl held his head up at four to five weeks, rolled over at four to five months, sat alone at five months, walked without support at nine months, and spoke his first word at eleven months. His language development may have been slightly delayed when evaluated at fourteen months. At that time gross motor function was normal.

Due to the finding of broad thumbs and fingers in other family members the possibility of Pfeiffer syndrome or Saethre-Chotzen syndrome was brought up. Apert syndrome was ruled out as no relative exhibited any form of syndactyly.

At the conclusion of the child's fourteen month exam the recommendation was made that he be re-examined in one year for possible hearing or visual problems. The family failed to follow up on this recommendation.

Darryl's next full physical is scheduled to occur in the summer of 1990 at The Children's Development and Rehabilitation Center, Portland Oregon.

Darryl had a comprehensive dental examination in June of 1989 at The Oregon Health Sciences University School of Dentistry, Department of Pediatric Dentistry. At that time he was noted to have the severe maxillary crowding which is often seen in Crouzon's syndrome. The crowding tended to center around the premolar-cuspid region, forcing the eruption of the maxillary cuspids to the buccal of the dental arch (See occlusion photos). His occlusion was found to be class one with an anterior cross bite of approximately one millimeter. The mandible appeared slightly small in size when compared to the maxilla.. No dental caries were noted but, several deep pits were present requiring sealants.

Darryl's midface was classic for Crouzon's, being rather hypoplastic in appearance. The eyes showed little evidence of proptosis. Other findings were within normal limits with the exception of the patient being somewhat overweight.

On June 11, 1990 Darryl was seen for comprehensive evaluation at The Oregon Health Sciences University's Child Development and Rehabilitation Center. One reason for this visit was to offer the family a definitive diagnosis of Darryl's condition. Other reasons the parents gave were the need for neurological and audiological follow up as recommended at the

time of Darryl's surgery.

At this exam Darryl was noted to be hyperactive. He is currently being treated for this condition with Ritalin. Also of note was a mild learning disability as reported by Darry's teachers. Darryl's hyperactivity and learning disability may be connected with his craniosynostosis. It would be very difficult to prove this either way as hyperactivity and learning disabilities are often related and are fairly common in the population at large. The only recommendation made concerning these problems was the consideration that Cylert be considered rather then Ritalin for the hyperactivity.

Psychologically, Darryl was found to be a pleasant young man who interacted well with the examining team. He tends to make friends easily and has no social problems other then his hyperactivity. In summary Darryl is a well adjusted twelve year boy.

Darryl's coordination was found to be lacking. He performed rapid alternating movements slowly and sloppily. He demonstrated a definite mild chorea of the outstretched hands.

An abnormal cranial shape and flattening of the face was found. No proptosis was noted. No visual abnormalities were found with uncorrected vision being 20/25. The audiological exam was normal.

It is interesting that while the final clinical impression given in the

craniofacial report is "Familial Craniosynostosis", most of the participents in Darryl's evaluation felt he had Saethre-Chotzen syndrome as opposed to his original diagnosis of Crouzon's syndrome. The problems found here in the diagnosis of this patient's syndrome point out the difficulties often encountered in differential diagnosis among the multitude of craniosynostotic syndromes.

This paper has examined the findings of Crouzon's syndrome in some detail. The different systems affected and the pathology noted have been described. Inheritance, psychological issues and surgical techniques were discussed. In addition other similar syndromes such as Apert and Saethre-Chotzen syndromes were compared and contrasted with Crouzon's syndrome where applicable. A case report of a surgically corrected patient diagnosed with Crouzon's syndrome was presented. It is hoped that the reader of this paper will now have a firmer grasp of Crouzon's syndrome, craniosynostosis in general, and the myriad of problems associated with these.

Feature	Apert syndrome	Crouzon's syndrome
Head	Hyperacrobrachycephalic	Brachycephalic
Facial Asymmetry	Yes	No
Proptosis	Less Severe	More Severe
Hypertelorism	Likely	No
Maxilla	Retruded, Hypoplastic	Same
Mandible	Relative prognathism	Same
Palate	Narrow w/ High Arch	Narrow
Cleft	Possible	Unlikely
Malocclusion	Severe Cl. 3	Less Severe Cl. 3
Nasopharynx	Short	Very Short
Skin	Seborrheic	Normal
Clavicle	Short	Normal
Limbs	Ankylosis	Normal
Hands	Bony Syndactylism	Normal
Intelligence	Often Retarded	

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⊘ = Deceased Female

2.3-2-Rev. 3–3/82

|□| = Adopted, in or out

= Identical Twins

FR = Fraternal Twins

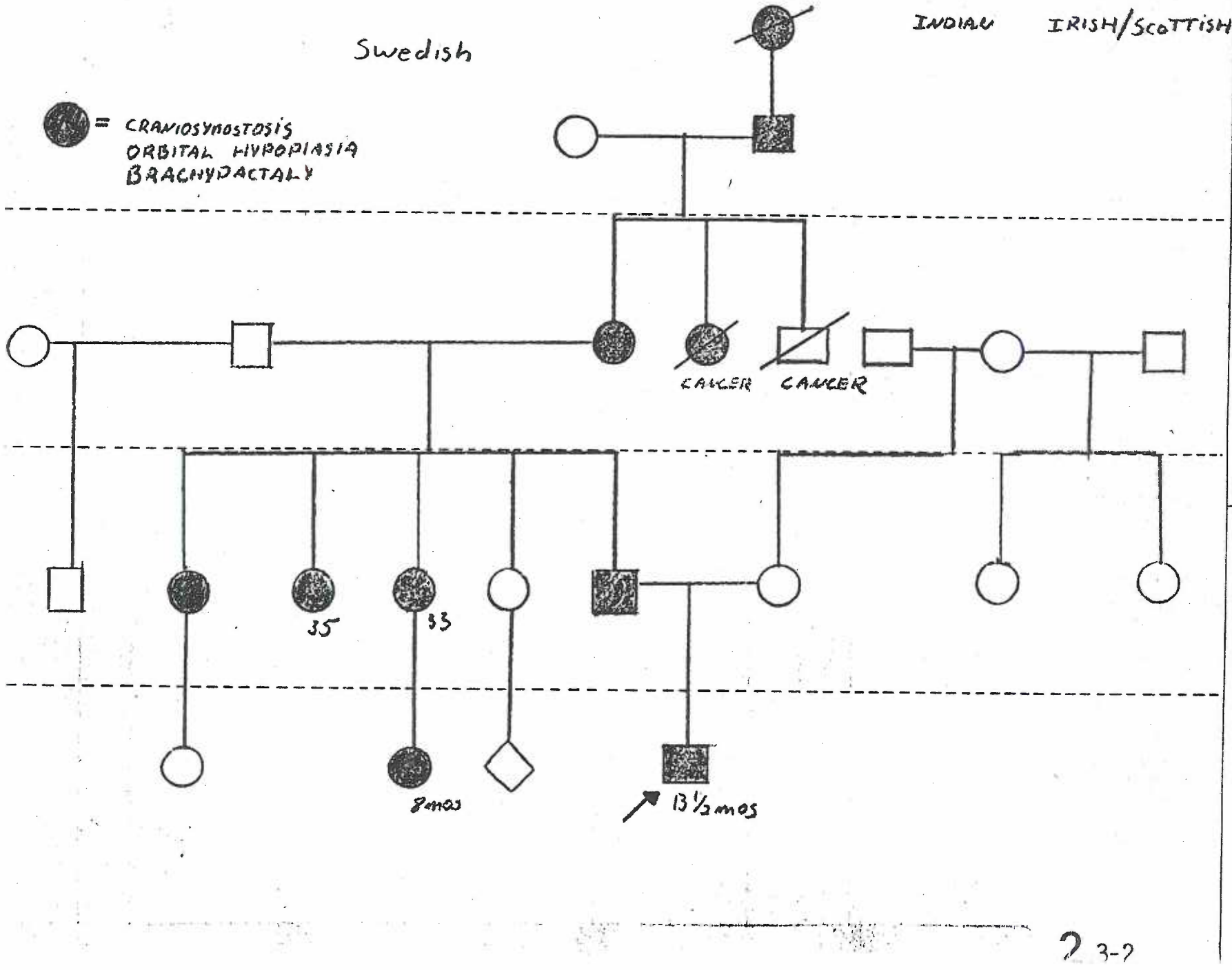
= Multiple Marriages

= Illegitimacy

= No Issue

◈ = Stillbirth

◇ = Miscarriage



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FAMILY HISTORY SHEET

Page No._____ Race (Paternal)_____

Family No._____ (Maternal)_____

Recorder_____ Religion_____

Date_____ Referring Diagnosis_____

Informant_____

Unit No.

Name

Birthdate

Date

Bldg.

Fl.

Rm.

2 3-2