Diagnosing Anencephaly in Archaeology: A Comparative Analysis of Nine Clinical Specimens from the Smithsonian Institution National Museum of Natural History, and One from the Archaeological Site of Kellis 2 Cemetery in Dakhleh Oasis, Egypt

by

Stevie L. Mathews
B.A. University of Central Florida, 2005

A thesis submitted in partial fulfillment of the requirements
for the degree of Master of Arts
in the Department of Anthropology
in the College of Sciences
at the University of Central Florida
Orlando, Florida

Summer Term
2008

Diagnosing Anencephaly in Archaeology: A Comparative Analysis of Nine Clinical Specimens from the Smithsonian Institution National Museum of Natural History, and One from the Archaeological Site of Kellis 2 Cemetery in Dakhleh Oasis, Egypt

In holdings at the University of Central Florida Library
Orlando Florida 32816

ABSTRACT

The inclusion of human fetal skeletons in the archaeological record can reveal much about past cultures' perception of life and death. The preservation of fetal remains in the archaeological record is a rarity, and the discovery of pathological skeletons is even rarer. A fetal skeleton from a Roman period cemetery (c. 31BC - 303AD) in the Dakhleh Oasis, Egypt, displays what are thought to be classic skeletal indicators of the neural tube defect, anencephaly. The published literature concerning the skeletal diagnosis of anencephaly is scant so in order to diagnose this individual it is pertinent to create a diagnostic standard. The purpose of this thesis is twofold – first to create a quantitative standard from which researchers can determine the presence of anencephaly in the archaeological record, thus ruling out trauma or taphonomic processes as reasons for missing cranial elements. The second objective of this research is to conduct a qualitative comparison in order to diagnose the individual from the Dakhleh Oasis. A comparative analysis of nine documented anencephalic skeletal remains housed at the Smithsonian Institute was conducted to create a diagnostic standard for the skeletal characteristics of anencephaly. The comparative analysis of the Dakhleh specimen supports the diagnosis of anencephaly.

.

To my ever supportive family

who have always encouraged me to follow my dreams

regardless of how outrageous they may seem.

ACKNOWLEDGMENTS

First and foremost, I would like to thank my family and friends for their encouragement and support over the course of my studies. I would also like to thank the faculty and staff of the anthropology department and specifically the members of my committee for their patience and guidance: Tosha Dupras, Matthew McIntyre, and Christopher Dudar. Furthermore, I am indebted to the Egyptian Supreme Council of Antiquities and members of the Dakhleh Oasis Project, in particular: Tony Mills, El Molto, and Peter Sheldrick. I am also thankful for the warm welcome and generous assistance from Dave Hunt and Don Ortner at the Smithsonian's National Museum of Natural History in Washington D.C. In addition, I am also grateful for the assistance provided by Sandra Wheeler and Lana Williams from the University of Western Ontario.

TABLE OF CONTENTS

LIST OF FIGURES

LIST OF TABLES

CHAPTER 1
GENERAL INTRODUCTION

During the 2003 winter field season an unusual fetus was excavated from grave number 563 in the Roman period Kellis 2 cemetery in the Dakhleh Oasis, Egypt as a part of the on-going research of the Dakhleh Oasis Project (Fairgrieve and Molto, 2000; Hope, 2001; Molto, 2001; Stewart et al., 2003; Tocheri et al., 2005; Dupras and Tocheri, 2007). It was not until the 2004 winter field season that the unusual nature of this individual was realized – something was missing. As this fetus' skeleton was laid out in anatomical position it became apparent that there was something wrong with all the cranial bones. Postcranially K2 B563 was complete and well preserved. It was at this time that it was speculated this fetus died as a result of a neural tube defect, more specifically, of anencephaly.

In order to find support for this hypothesis, the skeletal features of this fetus would need to be qualitatively compared to published literature as well as to other skeletons with this type of defect as well as a sample of non-pathological fetuses as a control. Unfortunately, there is very scant published evidence of the skeletal characteristics, archaeological or clinical, of anencephaly, which makes it nearly impossible to compare K2 B563 to published literature. Only one example of anencephaly found in an archaeological context has ever been published. This example, known as the Hermopolis mummy, was mentioned in several publications in the 1820s and once again in detail in 1847. Since its last mention the Hermopolis mummy has gone missing. However, the Smithsonian Institution's National Museum of Natural History (NMNH)

in Washington D.C. curates a large fetal skeletal collection which includes nine clinical cases of anencephaly which were donated to the collection in the 1800s and early 1900s (Huxley, 2005). These individuals span in age from approximately fifteen weeks gestation to full term. This collection makes it possible to conduct a diagnostic study of the cranial characteristics of anencephaly.

Because of the lack of published research on this topic, the purpose of this project was twofold. After ruling the possibility that the missing portions of the fetal skull were the result of decomposition, or that it had been damaged due to poor excavation technique (Miller and Simon, 2001) it is possible to say with certainty that the physical morphology of these cranial bones resulted from malformed growth during development. The first objective of this research was to devise standards that qualify what an anencephalic fetal skull looks like so that bioarchaeologists would be able to refer to while in the field should they think they have an anencephalic fetus present. Secondly, from the development of the diagnostic criteria, it then becomes possible to conduct a qualitative comparison of the K2 B563 individual from the Dakhleh Oasis.

Chapter 2 introduces neural tube defects through qualitative description so that criteria can be established for diagnosing anencephaly based on previously published literature. The chapter begins with an overview of early fetal growth and suspected causes for errors in the proper development of the neural tube. Although this paper is focused on anencephaly, the neural tube defect spina bifida is also discussed because of its high frequency of occurrence in association with anencephaly. Therefore, spina bifida and anencephaly and their corresponding subcategories are considered. The criteria used by the medical community for classifying which

type of subcategory is present within a given neural tube defect can vary. Thus certain defects, such as craniorachischisis, may be referred to as their own category in some sources, but may be classified within another. In this text craniorachischisis is considered to be a form of anencephaly.

Once a foundation of fetal development and origins of neural tube defects is established, an analysis of each cranial bone affected by anencephaly is presented. As the Hermopolis mummy from Egypt is the only published archaeological comparison (Miller and Simon, 2001; Saint-Hillaire, 1826) that can be made to K2 B563, an analysis of the scapulae, which are also affected by craniorachischisis, is included with the cranial elements. The pictorial representation of each anencephalic bone originated in a documented case of anencephaly and is shown in comparison to the same bone aged to the same gestational week of a non-pathological fetal skeleton. The sample of both nonpathological and anencephalic skeletons used in the comparison of K2 B563 for this study are housed at the NMNH. A description of the ossification of each bone included with the picture discusses details such as the timing of appearance of ossification center(s), the directional spread of bone growth, the week in which the bone ought to visually resemble adult morphology, as any other detail specific to that bone which may aid in the identification of an error due to a neural tube defect.

Chapter 3 builds upon the analysis of the individuals from the NMNH. Here, a further study reanalyzes all the skeletal elements from the NMNH in comparison to the suspected anencephalic fetus K2 B563 from the Dakhleh Oasis, Egpyt. A review of neural tube defects from chapter 2 is included as part of the introduction, as well as a discussion about Kellis 2. The

Hermopolis mummy is brought into context in order to be compared to K2 B563. The results of this section show pictures of each of K2 B563's malformed cranial bones next to the same bone of an anencephalic from the NMNH of the same age to further support conclusions made.

In chapter 4 the potential for further research is investigated. Also a comprehensive discussion of all materials and methods used for this thesis is reviewed. Finally conclusive evidence is reiterated for the classification of K2 B563 as an anencephalic individual.

References

Dupras TL and Tocheri M. 2007. Reconstructing infant weaning histories at Roman period Kellis, Egypt using stable isotope analysis of dentition. *American Journal of Physical Anthropology* 134:63-74.

Fairgrieve SI, and Molto JE. 2000. Cribra Orbitalia in Two Temporally Disjunct Population Sample from the Dakhleh Oasis, Egypt. *American Journal of Physical Anthropology* 111(3):319-332.

Geoffroy Saint-Hillaire E. 1826. Note about an Egyptian monster found in the ruins of Thebes, in Egypt, by M. Passalascqua. *Bulletin des Sciences Medicales* VII: 105-108.

Hope CA. 2001. Observations on the dating of occupation at Ismant el-Kharab. In *The Oasis Papers I: The Proceedings of the First Conference of the Dakhleh Oasis Project,* Marlow CA, Mills AJ (eds.) Oxbow: Oxford; 43-59.

Huxley AK. 2005. Gestational Age Discrepancies Due to Acquisition Artifact in the Forensic Fetal Osteology Collection at the National Museum of Natural History, Smithsonian Institution, USA. *American Journal of Forensic Medicine & Pathology*. 26(3):216-220.

Miller E, and Simon K. 2001. Anencephaly – Something Missing from the Archaeological Record? *Paleopathology Newsletter* 115:9-11.

Molto JE. 2001. The comparative skeletal biology and paleoepidemiology of the people from ‘ein Tirghi and Kellis, Dakhleh Oasis, Egypt. In *The Oasis Papers I: The Proceedings of the First Conference of the Dakhleh Oasis Project,* Marlow CA, Mills AJ (eds.) Oxbow: Oxford; 81-100.

Stewart JD, Molto JE, Reimer P. 2003. The chronology of Kellis 2: the interpretive significance of radiocarbon dating of human remains. In: *The Oasis Papers III: The Proceedings of the Third International Conference of the Dakhleh Oasis Project,* Bowen GE, Hope CA (eds.) Oxbow: Oxford; 345-364.

Tocheri M, Dupras TL, Sheldrick P, Molto JE. 2005. Roman Period Fetal Skeletons from the East Cemetery (Kellis 2) of Kellis, Egypt. *International Journal of Osteoarchaeology*. 15(5): 326.

CHAPTER 2
DIAGNOSING THE SKELETAL MANIFESTATIONS OF NEURAL TUBE DEFECTS WITH AN EMPHASIS ON ANENCEPHALY

Introduction

Neural tube defects are fairly uncommon in many modern populations due to advances in technology, medical care, and dietary improvements. It is known from historical records that birth defects of this nature were more frequent prior to industrialization, however little is known about infant mortality in the ancient world (Baker et al., 2005; Roberts and Manchester, 1995; Barnes, 1994). Often the only remaining evidence of birth defects exists in well preserved skeletons, and these conditions only exist in a few locations. In addition, it is an even a rarer occurrence if pathologic individuals were buried in the same location as others in their population, as many times these individuals were buried in other locations or not at all (Lewis, 2007; Parker Pearson, 1999).

In most paleopathological analyses it is common for archaeological specimens to be compared to clinical literature (Scheuer and Black 2000, 2004; Barnes, 1994) to reach a diagnosis. Unfortunately there is very little clinical literature pertaining to the skeletal manifestation of neural tube defects. When an individual is discovered in an archaeological context with a pathology such as anencephaly, the analysis and diagnosis becomes imperative for the reconstruction of past populations' ideology regarding the treatment of those afflicted with pathology. In order to diagnose the skeletal characteristics of a neural tube defect, such as

anencephaly, diagnostic criteria should be developed on a documented sample. Such a collection is housed at the Smithsonian Institution's National Museum of Natural History (NMNH) in Washington, D.C. The focus of this paper is to develop skeletal diagnostic criteria for anencephaly based on the NMNH collection.

Background of Neural Tube Defects with an Emphasis on Anencephaly

Neural tube (NT) defects are not known to have a single genetic or teratogenic cause, although a genetic basis is hypothesized (Shaffer et al., 1990) and teratogenic factors can often play a role in the development thereof (Bailey et al., 2003; Quinlivan and Gregory, 2003). Clinical consequences of NTDs can range from mild to lethal and are often associated with other defects (Lie, 2006). In 1991 the Center for Disease Control (CDC) put forth an effort to minimize the occurrence of NTDs by publicly recommending that women who were attempting to get pregnant to consume 400 µg of folate per day. This initiative stemmed from research that asserted that folate, a type of B-vitamin, played a crucial role in the closure of the neural tube (Hibbard and Smithells, 1968) coupled with clinical studies that monitored the health of expectant mothers, which were able to confirm the validity of the previously established folate research (Williams et al., 2002).

The intended effect of the CDC's recommendation was a decline by 50% in the occurrence of neural tube defects. However, an alarming number of women failed to follow these guidelines (Mills and England: 2001). In 1992, the U.S. Public Health Service made a similar public recommendation that all women capable of becoming pregnant ought to follow the same

guidelines as those who were in the high risk category. Both of these recommendations were based on cases studies showing that daily consumption of 400 µg of folic acid significantly reduced the risk of pregnancies with a neural tube defect (Erickson, 2002). Although the positive effects of increased levels of blood folate were made known, there was little voluntary compliance with these standards (Daly and Mills, 1997).

Just like the fortification of dairy products with vitamin-D to prevents rickets, scientific evidence confirmed that folic acid supplements taken periconceptionally could – and did –reduce the risk of neural tube defects (Mathews et al., 2002). These results have been so reliable that a series of worldwide public health policies have been established. In March of 1996 the United States' Food and Drug Administration (FDA) made the recommendation that all cereal grains products be fortified with folic acid at the level of 140 µg/100 grams of grain. This recommendation became a nationally recognized mandatory requirement in January of 1998. Foods that were folic acid fortified in addition to having already been enriched with iron, thiamin, riboflavin, and niacin include flour, rice, breads, rolls and buns, pasta, corn grits, corn meal, farina, macaroni, and noodle products.

It should also be noted that research has shown (Shane, 2003) that the fortification of foods with folate may actually be harmful to pregnant women, and that the appropriate level of folate may be less than what is consumed on average (Ulrich and Potter, 2006). Therefore it is suggested that controlled field trials need to continue in order to ascertain whether or not folic acid fortification actually benefits both the pregnant woman and her unborn child (Wharton and Booth, 2001).

Physicians and scientists now need to develop a better understanding of how many cases of NTDs are the result of inadequate folic acid consumption alone, and how many develop from other causes that are not related to levels of folic acid. There is still a need for further research to better understand the biological mechanisms that prevent NTDs based on folate levels, and even more so the origin of cases that have no connection to folic acid consumption.

Regardless of cause, the improper formation of the embryonic neural tube during the third and fourth week of development results in defects of the skull or of the spine, and sometimes in both. This is the definitive point in time when a number of severe cranial and spinal defects often develop. Generally, neural tube defects can be described as life threatening developmental abnormalities of the central nervous system. Although severe, neural tube defects are among the most common out of a range of congenital malformations in humans (Green and Copp, 2006), yet the variation among these types is poorly understood (Moore , 2006).

It is important to note that due to the severity of most neural tube defects, many pregnancies will spontaneous abort prior to the mother's knowledge of pregnancy, or prior to the medical recognition of this defect (Barnes, 1994). It is for this reason that it is nearly impossible to estimate the rate of total occurrence for neural tube defects. in a publication by Lie (2006) the author suggests that prevalence rates can be estimated post 16 weeks gestation – presuming all cases among pregnancies are correctly diagnosed and counted. Although this may be possible, any attempt to base the rate of incidence on cases that survived to late term or even birth would be dramatically skewed because of the lethality of these defects. In addition, the common

decision to electively terminate the pregnancy when carrying a child with a severe neural tube defect also decreases the ability to accurately assess the incidence of this defect.

To discuss the etiology of NTDs it is essential to develop an understanding of fetal growth patterns, as the knowledge of how bone develops in utero is critical to comprehending how and when this type of developmental error occurs. The human embryo develops rapidly after conception. The embryonic period, also called organogenesis, occurs from the third to eighth week of development (Sadler, 2006). During the third and forth weeks of organogenesis the accurate formation of the neural tube is of the utmost importance in the proper development of a human embryo.

At the beginning of the third week of gestation the primitive streak forms along the midline of the ovular shaped bilaminar germ disc (Larsen 2001). The formation of the primitive streak is followed by a deepening and elongation of this furrow thus forming the primitive groove, which is surrounded by the primitive node. The development of this structure is meant to ensure bilateral symmetry in the embryo, as well as giving rise to the endoderm and mesoderm for the head, the trunk, and the tail of the body.

With the progression of the primitive node comes the onset of the formation of the axial skeleton. This development begins with gastrulation (Larsen, 2001), which is the process where the cellular structures for the endoderm and mesoderm develop. At the same time the primitive groove stretches and undergoes a compositional change. This tube-like structure becomes the notochord, which is responsible for forming the spinal cord and vertebral bodies. As gastrulation

continues, structures develop that provide the framework for the brain, vertebral column, basioccipital, and sphenoid (Sadler, 2006).

One of the crucial events responsible for proper development is the conversion of the neural plate into the neural tube. As this primitive structure thickens and becomes more complex, it gives rise to the neural tube. The formation of the neural tube is the first step in the development of the central nervous system. The cranial end closes between 20 and 22 days after conception, whereas the caudal end closes between 22 and 26 days after conception (Barnes, 1994). Errors at this stage often prevent closure of the neural tube thus leading to a defect. Abnormalities of the neural tube closure not only affect the development of the central nervous system but also interfere with vertebral arch and cranial vault morphogenesis (Larsen, 2001).

The most common NTDs are spina bifida and anencephaly. Both of these categories contain several subcategories classified according to severity. Unfortunately, data regarding the specific subcategory of either spina bifida or anencephaly is generally not recorded, therefore little is known about the potential demographic differences between the types. Spina bifida ranges in severity from resulting in fatality, to living with the defect unknowingly. Anencephaly is equally fatal regardless of severity. Most anencephalic fetuses result in a premature birth. Of those that survive to full-term, nearly all individuals die within a few hours, or days at most, after being born (Dambska and Wisniewski, 1999).

Spina Bifida

This most common of NTDs is spina bifida (Lie, 2006). Spina bifida, as a term however, refers to a series of defects which range in severity. This defect is caused by a spinal dysraphism, or failure of the neural tube to close at the caudal, or posterior, end of the neuropore. Spina bifida is caused by a direct disruption in the overlying vertebral arches in that they remain underdeveloped, therefore the lamina do not fuse along the midline, resulting in an open vertebral canal.

There are three types of this defect: spina bifida occulta, spina bifida cystica (also referred to as myelomeningocele), and meningocele. The difference between the types is rooted in the severity of each. Spina bifida occulta is classified by the failure of the vertebral arch of one vertebra to fuse to the other. In this case the underlying neural tube develops normally, remaining in the vertebral canal. Thus, an occulta defect can be rather mild and can be surgically corrected. Often the mildest cases of spina bifida occulta go undetected as its root in Latin, *hidden*, suggests. The outward appearance of this defect along the spine is usually skin-covered as normal. Other variations include a dimple in the skin, a benign tumor called a lipoma, or even a small patch of hair directly over the unfused vertebra (Lemire, 1988).

Spina bifida cystica, the more severe form, exhibits the same skeletal manifestation as spina bifida occulta in that the vertebral arches fail to meet and fuse at the midline. The origin for this defect is caused by an abnormality that occurs during secondary neurulation and can occur at any single vertebra, but is most common in the lower lumbar or sacral region (Green and Copp, 2006). This category of spina bifida is considered more severe because the unprotected spinal

cord protrudes through the unfused bone resulting in its exposure outside of the body. The spinal cord may or may not be enclosed in a sac filled with cerebrospinal fluid. Although a more severe form of spina bifida, scientific advances in medicine now allow this developmental error to be surgically corrected through a hysterotomy, or while the fetus is still in the mother's uterus (Walsh and Adzick, 2003).

Meningocele, also known as myeloschisis or myelocele, was formerly grouped with spina bifida cystica. This defect is rare in comparison to the other two types of spina bifida, and most often results in fatality due to its extreme severity. In this form, the vertebral arches also fail to meet and fuse at the midline, but the difference lies in that the spinal cord develops normally and remains within the vertebral canal. The life threatening abnormality results from the meninges, the membranes that envelop and protect the central nervous system, being pushed through the unfused vertebrae resulting in a sac filled with cerebrospinal fluid. Developmental pressures can force this sac outside of the body, as in spina bifida cystica, or into the retroperitoneum, which is the anatomical term for the space behind the abdominal cavity (Moore, 2006).

Individuals with more severe cases of spina bifida, around 50% of those that go undetected and untreated, die around the time of birth (Lie, 2006). Other more severe neural tube defects, such as anencephaly and craniorachischisis, also occur when the neural folds fail to fuse, as well as fail to separate from the ectoderm during primary neurulation. Defects this serious most often result in spontaneous abortion; those fetuses carried to term invariably die within hours or days after birth.

Anencephaly

Anencephaly is a neural tube defect in which the cranial or anterior end of the neuropore remains open and is clinically classified into three categories based on severity: meroanencephaly, holoanencephaly and craniorachischisis. In most cases when the term anencephaly is used it is meant to refer to meroanencephaly. The morphology of this manifestation of anencephaly is a lack of cranial vault bones, which results in an exposed dorsal mass of undifferentiated neural tissue (Isada et al., 1993). Much of the brain is underdeveloped or absent altogether. Typically, the brainstem and portions of the midbrain are all that are present; the cerebral hemispheres do not develop. The pituitary gland is hypoplastic or absent (Moore, 2006). The portion of the brain that is present controls basic breathing, cardiovascular, suckling and elimination reflexes; therefore the infant will never gain consciousness or feel pain (Merker, 2007).

Holoanencephaly is more severe. The physical manifestation of this type of anencephaly is seen in the bony defect extending from the vault through to the foramen magnum (Dambska and Wisniewski, 1999) in which the posterior aspect of the skull remains entirely open. Holoanencephaly is commonly mistaken to be the same defect as acephaly. Acephalic individuals completely lack a cranium and brain.

Craniorachischisis, the third type of anencephaly, is not always classified with anencephaly, although these fetuses lack cranial bones and brain tissue. Craniorachischisis manifests as the combination of both holoanencephalic characteristics and continuous spina

bifida in one individual. This sometimes warrants craniorachischisis to be classified as a unique defect. Beyond the cranial malformations seen in anencephaly, individuals with craniorachischisis can also develop deformed vertebrae, scapulae, clavicles, and ribs. For the purpose of this paper craniorachischisis is classified as the most severe form of anencephaly. This condition is extremely rare, although it occurs in a higher frequency among groups with higher neural tube defect rates, such as the United Kingdom and Northern China (Dolk et al., 1991; Moore et al., 1997).

Anencephaly and craniorachischisis both result from the failure of the neural folds to fuse during the third and fourth week after conception. Hysterotomic techniques that can correct some forms of spina bifida, do not possess the capacity to correct anencephaly or craniorachischisis and both types are equally fatal, regardless of severity. It seems unlikely that this type of surgical procedure will ever be able to correct these defects because these individuals do not develop brain matter very early in pregnancy. Thus, a procedure would have to be developed that would be able to correct this problem before most women are even aware that they are pregnant.

Previous Research Studies of Anencephalic Skeletons

The beginning of scientific theory regarding the development of a fetus was first recorded in ancient Egypt within the Kahun papyrus. This tradition of scientific inquiry continued from Egypt into study by Greek and Roman philosophers. Pliny the Elder was one of the first to write a portrayal of the appearance of neural tube defects (Charon, 2005). It was not until the medical

advancements in the Renaissance that any physician attempted to describe and classify what we now refer to as neural tube defects.

The man responsible for coining the term *anencephaly* is Étienne Geoffroy Saint-Hillaire. Saint-Hillaire (1772-1844), a French naturalist and colleague of Jean-Baptiste Lamarck, spent most of his life devoted to studying embryology and the comparative anatomy of humans and non-human primates. In 1798, Saint-Hillaire was selected as one of 167 scientists on Napoleon's great scientific expedition to Egypt. During his three years in Egypt he met and befriended Joseph Passalacqua, a collector of Egyptian mummified animals. During Passalacqua's travels he acquired a collection of sacred mummified baboons from Hermopolis in Western Thebes, known today as Dra Abu el-Naga. After returning to France Saint-Hillaire examined Passalacqua's collection of mummified baboons. To his surprise, amongst the collection he discovered an anencephalic fetus, which he recognized immediately as human due to his extensive research on the subject. Saint-Hillaire published numerous descriptions of the Hermopolis mummy during the 1820s. After his death in 1844 his son, Isidore Geoffroy Saint-Hillaire, continued his father's research on the Hermopolis mummy and anencephaly in general. He published his own description of the mummy in 1847 along with 2 plates showing a detailed view of the Hermopolis mummy from the front, side, and rear.

Since the mid-1800s numerous advances in medical technology allowed for the advancement of in depth studies into embryology. Although anencephaly is a neural tube defect that has continued to manifest in fetuses since the earliest records first annotate, very little research has been published. Early investigations of abnormal development focused on studies of

epidemiology aimed at etiology (Stevenson et al., 1966; Naggan and MacMahon, 1967). While other studies centered on anatomic and embryologic morphology (Abd-El-Malek, 1957; Keen 1963; Marin-Padilla, 1965) and pathogenesis in animals (Giroud 1960; Langman and Welch, 1966). In 1978, a new approach was attempted which compared craniofacial skeletal development in normal and anencephalic fetuses. The findings of this study were written as a series of three publications that discuss the skeletal manifestations of anencephaly (see also Garol et al., 1978; Metzner et al., 1978). Unfortunately the aging technique used for the fetuses at the time, length of the foot, is no longer deemed reliable; therefore the discussion about the age in gestational weeks for these individuals has been excluded.

The first of the three articles (Fields et al., 1978) analyzes the morphology of the cranial base, whereas the second (Garol et al., 1978) focuses on the calvarium, and the third (Metzner et al., 1978) observes the facial skeleton. These studies used anatomy, radiography, and histology to analyze the morphology of anencephaly from an unknown collection. For their research, twelve anencephalics were selected out of 28 available specimens ranging from 26 to 40 weeks gestational age. The only information provided regarding the origin of these anencephalic individuals states that they were previously preserved in 10% formalin, suggesting they were curated in some way.

For analysis the 12 anencephalics of varying severity were “decapitated” and examined, photographed, and radiographed before the soft tissue was dissected (Fields et al., 1978) and the periosteum was left intact to hold the bones in position after drying. After dissection the skulls were “divided in a parasagittal plane to preserve the midline structures” (Fields et al., 1978:57).

Although the term dissected was used, there is no account of the soft tissue being analyzed during this study. Even though the title of the first article states that Fields and colleagues were analyzing the cranial bases of these 12 anencephalics, pages 58-60 describe the configuration of the cranial floor; the difference between these two terms is not clearly defined in the article and is left up to the reader to infer based on the images. It is unfortunate that the postcranial elements were not analyzed along with the crania.

Unfortunately, the descriptions provided by Fields et al. (1978) are less about the morphology of each cranial element and are more about angles and the shape of cranial structures as a whole. Prior to this study, it was well known that the cranial base of a normal fetus was ovoid in shape. This study found that meroanencephlics' cranial base was trapezoidal in shape and the cranial base of holoanencephalics is triangular (Figure 2.1). Although the cranial

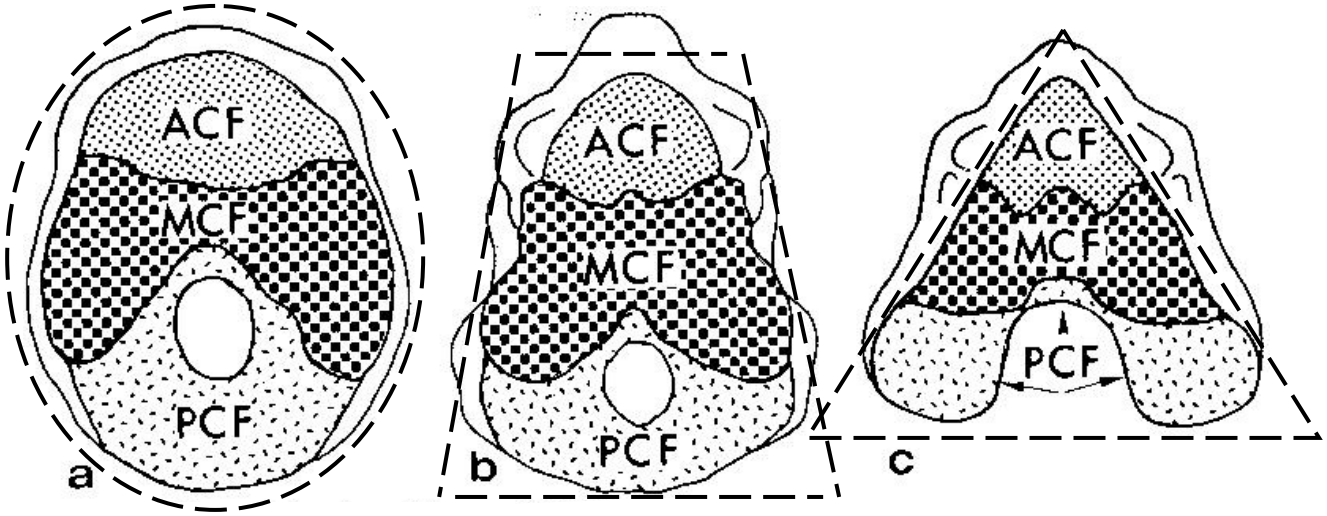


Figure 2.1: Schematic drawings of the cranial base in a normal fetus (a), in a fetus with meroanencephaly (b), and in a fetus with holoanencephaly (c). Note: the shape of the non-pathological fetus (a) is annotated by a dashed oval, of the meroanencephalic by a dashed trapezoid, and of the holoanencephalic (c) by a dashed triangle respective to the shape they actually display. The original author's abbreviations stand for anterior cranial fossa (ACF), middle cranial fossa (MCF), and posterior cranial fossa (PCF). *Source: Fields et al., 1978.*

bones were not described individually, the drawings of the cranial floors indicated what can be expected of each cranial element in both meroanencephaly and holoanencephaly as compared to normal fetuses based on proportion (Figure 2.2). Fields et al. (1978:61) make a general observation about the malformed sphenoid, stating that the, "greater wings were reduced in their anterior and lateral vertical development, which in turn reduced the width of the anterior portion of the middle cranial fossa". The authors also note that the sphenoid's articulation with several other cranial bones, or lack thereof, must have caused further malformations.

Garol et al. (1978) discusses the aperture that results from anencephaly and the variation that can accompany each form. In meroanencephalics the calvarial defect is described as ranging from, "…one to several centimeters in diameter, the latter exposing the entire cranial floor" (Garol et al., 1978:67). It was also noted that as the size of the defect increased, "…the size, shape, and spatial orientation of the calvarial bones were more severely altered relative to

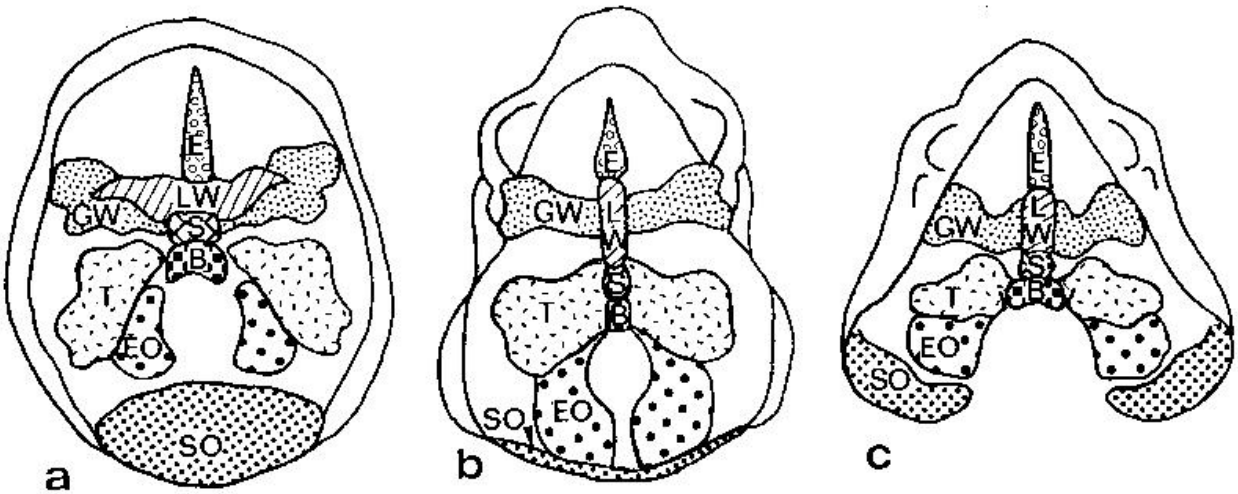


Figure 2.2: Schematic drawings of the cranial floor in a normal fetus (a), in a fetus with meroanencephaly (b), and in a fetus with holoanencephaly (c). *Source: Fields et al., 1978.*

Note: the original author's abbreviations stand for basiocciput (B), ethmoid (E), exoccipital (EO), greater wing of the sphenoid (GW), lesser wing of the sphenoid (LW), petrous temporal (T), sphenoid (S), supra-occipital (SO), and vomer (V).

the normal" (Garol et al., 1978:68). This observation is very valuable to researchers who may analyze disarticulated anencephalic fetal cranial elements.

The discussion of the defect in each type of anencephaly in Garol and colleagues' (1978) article includes a more inclusive morphological description of the bones affected. It seems that the less severe of the meroanencephalics had a nearly normal calvarial shape. This may suggest that they were misclassified as anencephalic, or that there is a far less severe form of anencephaly not discussed in modern clinical literature. In those with a more extensive defect the, "parietals were absent. The frontal bones diverged laterally anterior to the crista galli and extended posterolaterally until they approximated the squamous temporal bone. The occipital bone assumed a nearly horizontal orientation" (Garol et al., 1978:68).

In the study by Garol and colleagues, (1978) the skeletal morphology of holoanencephaly and craniorachischisis is described. The discussion of the holoanencephalics and those with craniorachischisis fall more in line with the classic descriptions of anencephaly. The cranial structures posterior to the foramen magnum are described as diverging laterally, and the squamous portions of the bones were markedly reduced in size, as well as a total absence of the parietal bones. The interparietal portion of the occipital has been relocated laterally and resembles a wing that trails off of the supra-occipital. The individuals with craniorachischisis displayed the same missing cranial elements, with a similar resemblance in defect with the exception of a greater width of opening in the posterior of the skull. A comparative sketch of the affected bones arranged by the varying severities of anencephaly is included. Figure 2.3 shows a superior view of the cranial elements, and Figure 2.4 is a side view of the same analysis. Fields

and colleagues concluded that, “the morphological alterations of the anencephalic cranial floor are due to the altered size, form, or duration of the neural functional matrix, which in turn influences adaptive changes in the membranous bones of the neurocranium and the facial skeleton” (1978:64).

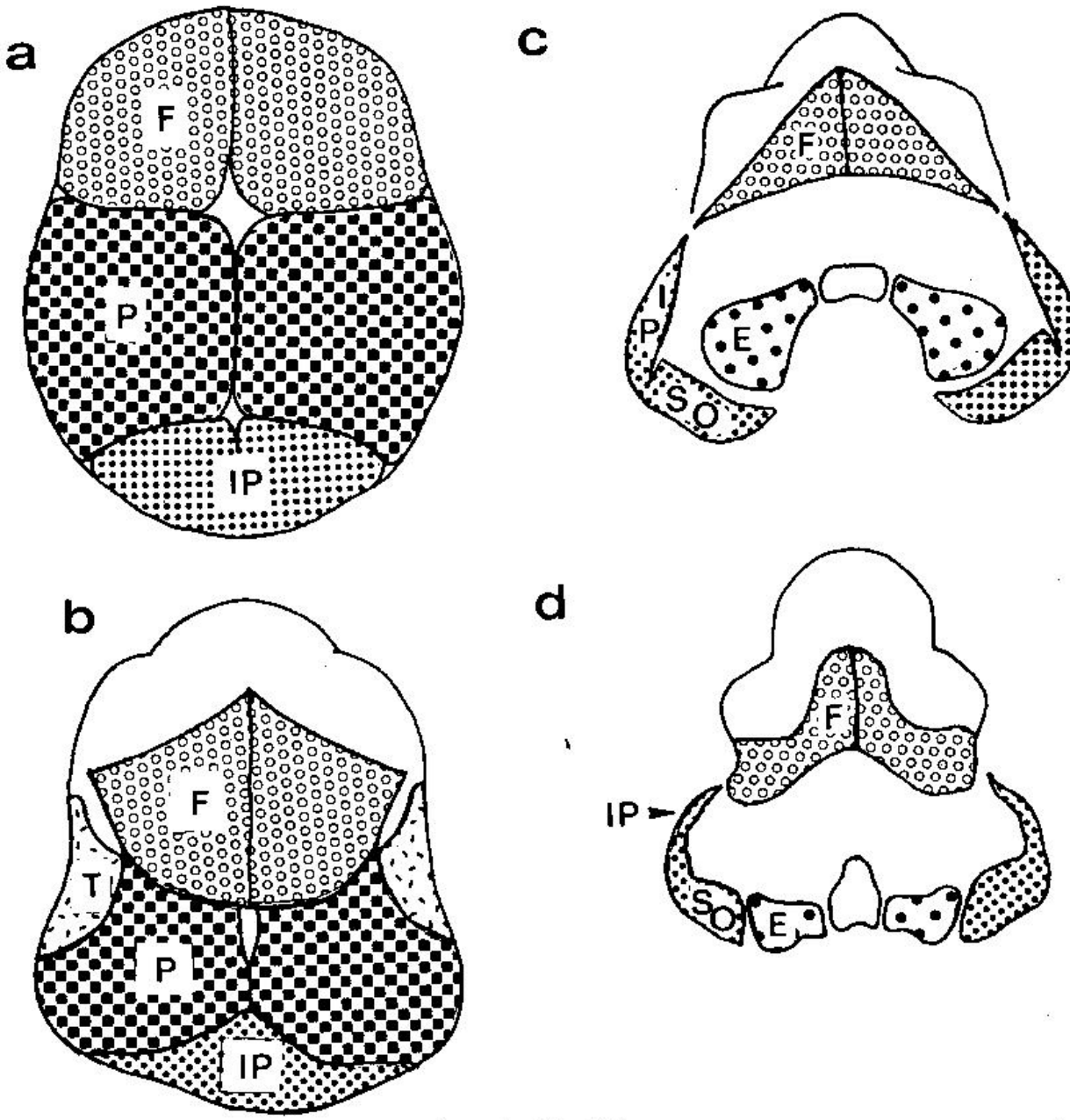


Figure 2.3: Tracings made from basilar radiographs. The sketching is of a normal fetus (a), a fetus with meroanencephaly (b), a fetus with holoanencephaly (c), and a fetus with craniorachischisis (d). *Source: Garol et al., 1978.*

Note: the original author's abbreviations stand for exoccipital portion of the occipital bone (E), frontal bone (F), interparietal portion of the occipital bone (IP), parietal bone (P), supra-occipital (SO), and temporal bone (T).

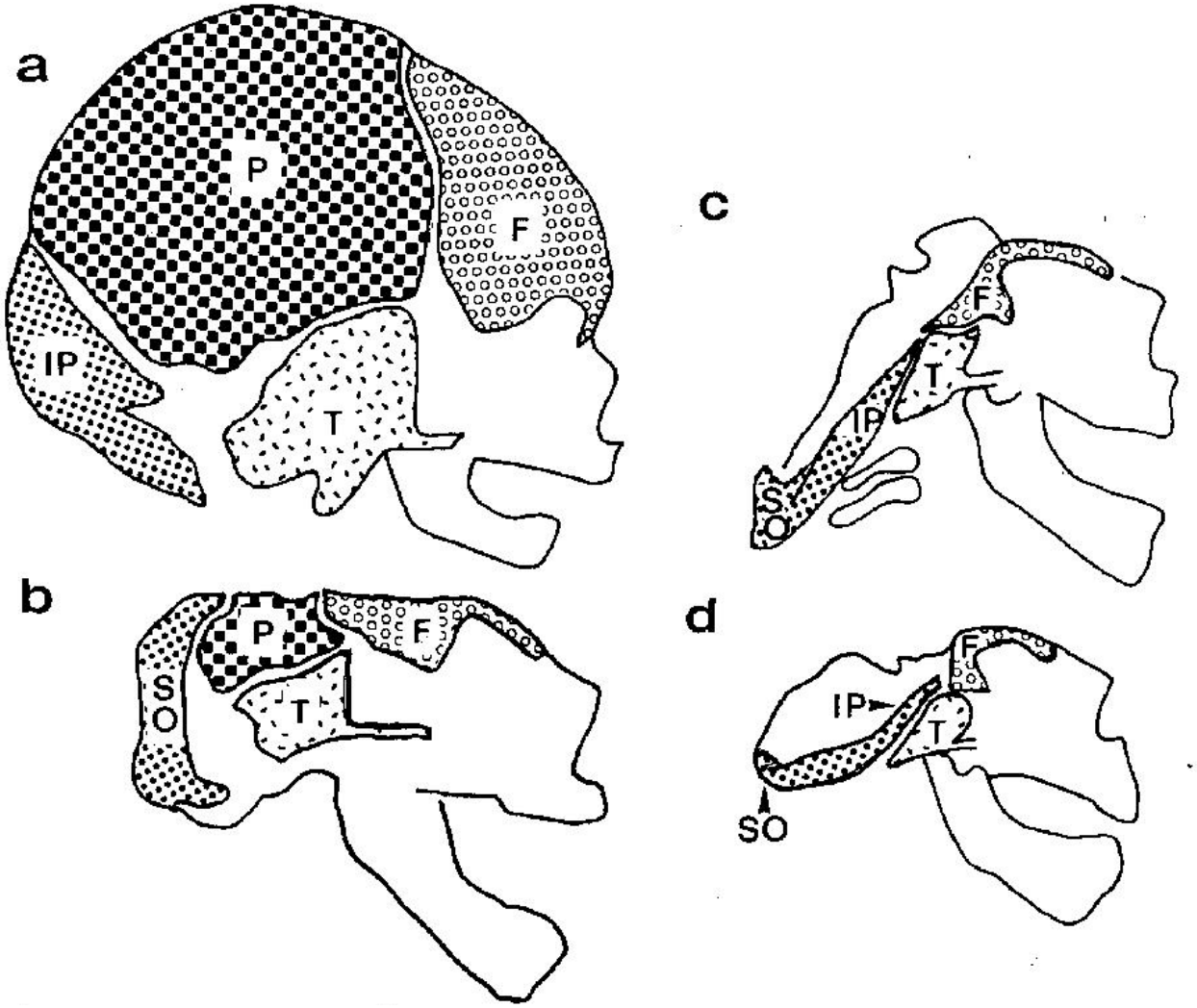


Figure 2.4: Tracings made from lateral radiographs. The sketching is of a normal fetus (a), a fetus with meroanencephaly (b), a fetus with holoanencephaly (c), and a fetus with craniorachischisis (d). *Source: Garol et al., 1978.*

Note: the original author's abbreviations stand for exoccipital portion of the occipital bone (E), frontal bone (F), interparietal portion of the occipital bone (IP), parietal bone (P), supra-occipital (SO), and temporal bone (T).

Bone Growth Affected by Neural Tube Defects

The earliest recorded appearance of osseous growth is used to define the beginning stage of that particular bone. It is important to note, however, that due to natural human variation, stages of ossification may begin one to two weeks gestation after the timing that is listed. Also in rare situations where the skeleton is malformed due to a NTD the timing for the appearance of ossification centers and fusion of certain boney elements are often disrupted. Detecting and properly diagnosing a developmental defect such as anencephaly in an ancient population can reveal not only something about the individual that died, but also about the culture from which it came. The following discussion covers the cranial elements affected by the NTD anencephaly. Although not all forms of anencephaly affect the morphology of the scapulae, it does occur with craniorachischisis, therefore they are included along with the cranial bones.

Parietals

The parietals form the majority of the left and right side walls of the brain case (Figure 2.5). They articulate with one another, with the squamous portion of the occipital, with the superior border of the temporals, with the greater wings of sphenoid, and with the frontals. During normal fetal growth several centers of ossification will appear, spreading rapidly across the bone. Ossification of the parietal begins in what will become the parietal eminence during the 7th week of intra-uterine growth (Scheuer and Black 2000, 2004). Bone growth spreads outward very rapidly as the parietal resembles the adult shape as early as 14 weeks (Baker et al.,

2005). From this point both parietals expand at a fairly continuous rate increasing in size and thickness until the fetus has reached full term.

Figure 2.5: Superior view of an articulated, but unfused, fetal cranium (23 weeks gestation). The parietals are indicated by arrows.

Occipital

The occipital bone is a very dense bone which functions to protect the brain. It makes up the base and rear of the cranium. Several centers of ossification appear early during development and bone growth spreads rapidly, developing into four parts: the squamous portion, the pars laterialis and the basion (Figure 2.6). Under normal conditions these four components should fuse during fetal growth medially and at the sutura mendosa. The squamous portion of the occipital articulates with the parietals, all four parts articulate with the temporals, and the basion articulates with the sphenoid.

Ossification of the occipital to begins during the 8th week of intra-uterine life when ossification centers for the pars interparietalis and pars laterialis first appear (Scheuer and Black 2000, 2004). The pars interparietalis form out of mesenchymal tissue, whereas the pars lateralis form endochondrally. The following week ossification of the pars supra-occipitalis begins in a cartilaginous precursor. The basion beings ossification during the 11th gestational week (Scheuer and Black 2000, 2004).

The supra-occipital and interparietal parts begin to fuse together about halfway through the third month starting in the middle of the bone, yet retaining a vascular channel called the suture mendosa. Bone growth spreads fairly rapidly through these bones uniting the two halves of the pars interparietalis together as well as the halves of the supra occipitalis. By the 5th month fusion is complete resulting in a unified squama. The pars lateralis and the basion begin to fuse to the squamous portion of the occipital during fetal growth but are not completely fused until the age of 3, and sometimes as late as six. These structures form the foramen magnum which

allows the spinal cord to pass from the brain down the spinal column. Complete fusion of the occipital does not occur until the age of 6, and sometimes as late as 8 (Baker et al., 2005).

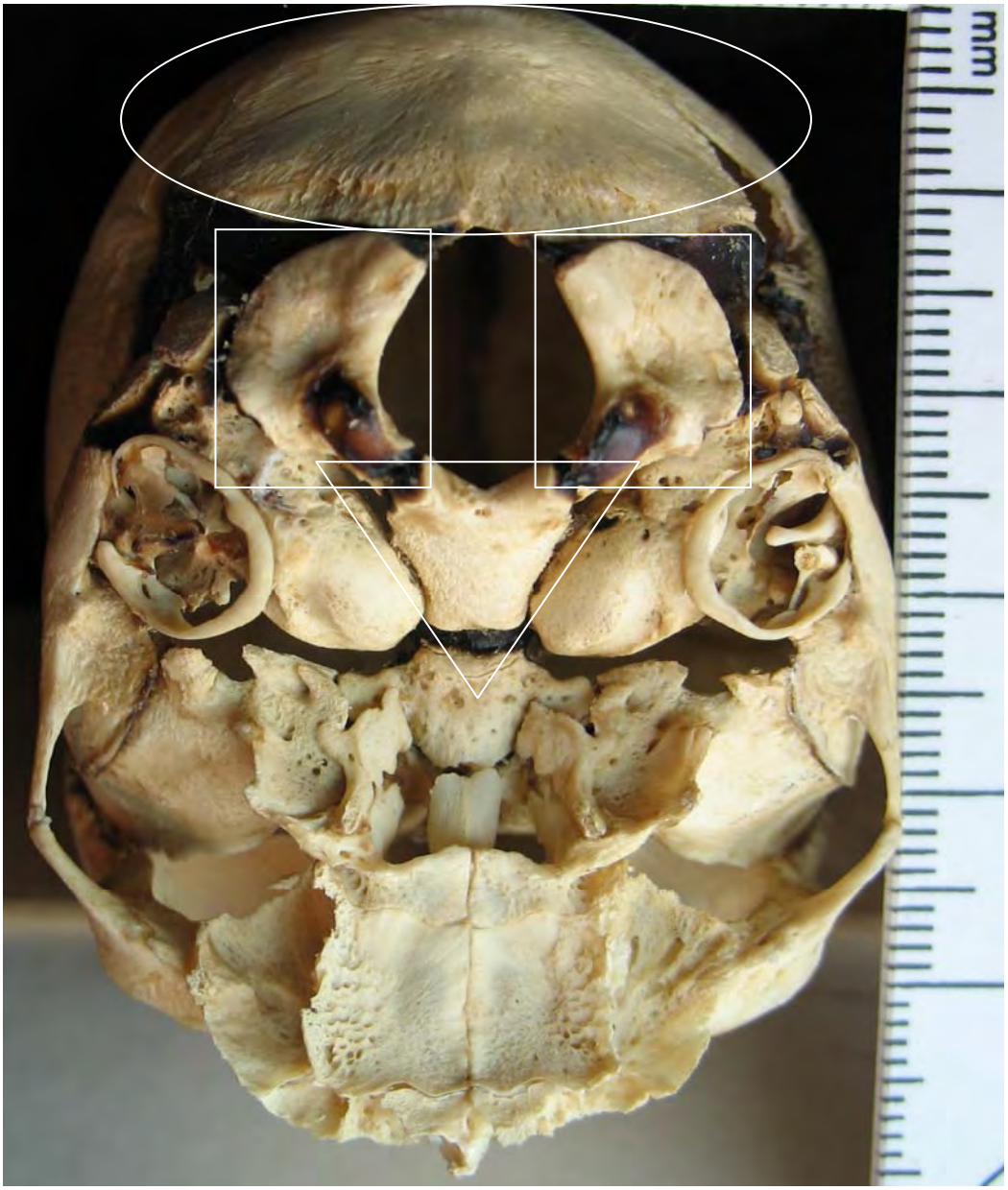


Figure 2.6: Inferior view of an articulated, but unfused, fetal cranium (23 weeks gestation). The squamous portion is indicated with an oval, the lateral parts are indicated by squares, and the basion is indicated by a triangle.

Temporals

Like the occipital, each temporal is also a dense bone. These bones, one on either side of the head, house the structures essential for hearing and maintaining balance. They partially make up the base of the skull and the lateral walls of the cranial vault (Figure 2.7). The squamous portion of the temporal articulates with the parietals, the greater wing of the sphenoid and the zygomatics. The petrous portion articulates with the occipital, with the mandible, and with the sphenoid. During normal fetal growth several centers of ossification will appear, developing into the petromastoid (often referred to as the petrous portion), squamous, tympanic ring and styloid process. During the final weeks of fetal development the tympanic ring fuses to the squamous portion becoming the squamotympanic.

Prior to unification the temporal develops three main parts: a squamous portion, a petrous portion, and the tympanic ring (Figure 2.8). The temporals also house the auditory ossicles; the incus, stapes, and malleus (Figure 2.9). Under normal conditions the three main components should begin to fuse during late fetal growth. Fusion completes during the first year of life when the petrous and squamous fully fuse into a single bone.

Ossification of the temporal begins with the squamous portion around 7 weeks gestation (Baker et al., 2005). Typically, only one center appears at the base of the zygomatic process. From this point ossification spreads from the mesenchymal tissue posteriorly to form the squama. At about 9 weeks the primary ossification center for the tympanic portions appear (Scheuer and Black 2000, 2004). Within four weeks ossification has advanced far enough for the bone to resemble the ring shape.

The ossification of the petrous portion begins during the 16th week of intra-uterine life. This bone forms endochondrally with the emergence of up to fourteen separate ossification centers. Once ossification begins it progresses fairly rapidly and is completely ossified by week 23 (Scheuer and Black 2000, 2004). During the 34th week the squamous, the tympanic ring, and the petrous portion begin to fuse into a single element.

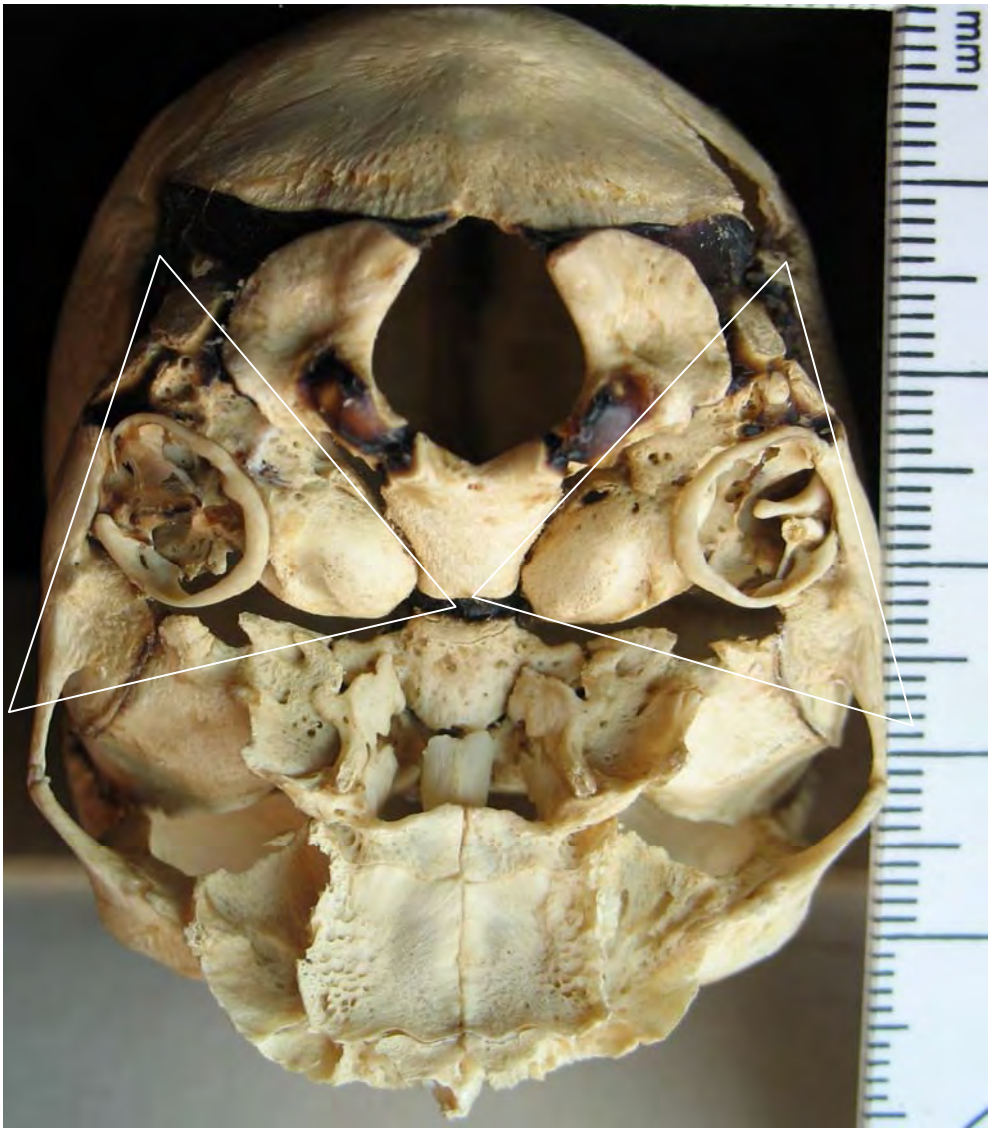


Figure 2.7: Inferior view of an articulated, but unfused, fetal cranium (23 weeks gestation). The temporals are indicated by triangles.

Figure 2.8: (Left side) Disarticulated fetal temporal (32 weeks gestation) displaying the squamous portion (top), the petrous portion (middle) and the tympanic ring (bottom) prior to fusion.

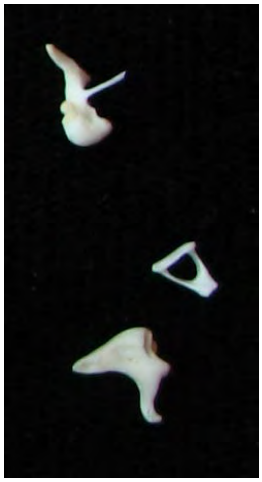


Figure 2.9: Fetal auditory ossicles (32 weeks gestation): malleus (top), stapes (middle) and incus (bottom).

Frontals

The frontals, comprised of a right and a left half, make up the superior portion of the eye orbit, and the forehead region which protects the brain (Figure 2.10). They articulate with the parietals, greater wings of the sphenoid, zygomatics, maxillae, lacrimals, nasals, and ethmoid.

Each half of the bone ossifies from a single center beginning in the 6th week gestation (Scheuer and Black 2000, 2004). This center forms intramembranously in what will become the anterior section of the orbital plate. Ossification spreads rapidly during the 7th and into the 8th week through the squamous portion of the frontal and then through the orbital portion (Baker et al., 2005). At 10 weeks gestation the supercilliary arch and zygomatic processes also begin to develop. By 13 weeks ossification of the orbital plate is complete (Scheuer and Black 2000, 2004).

The frontal is one of the earliest bones to become recognizable in the fetal skeleton due to its early ossification. Although both halves form relatively early and ossification spreads rapid throughout the bone, they do not fuse into a single bone until after birth.

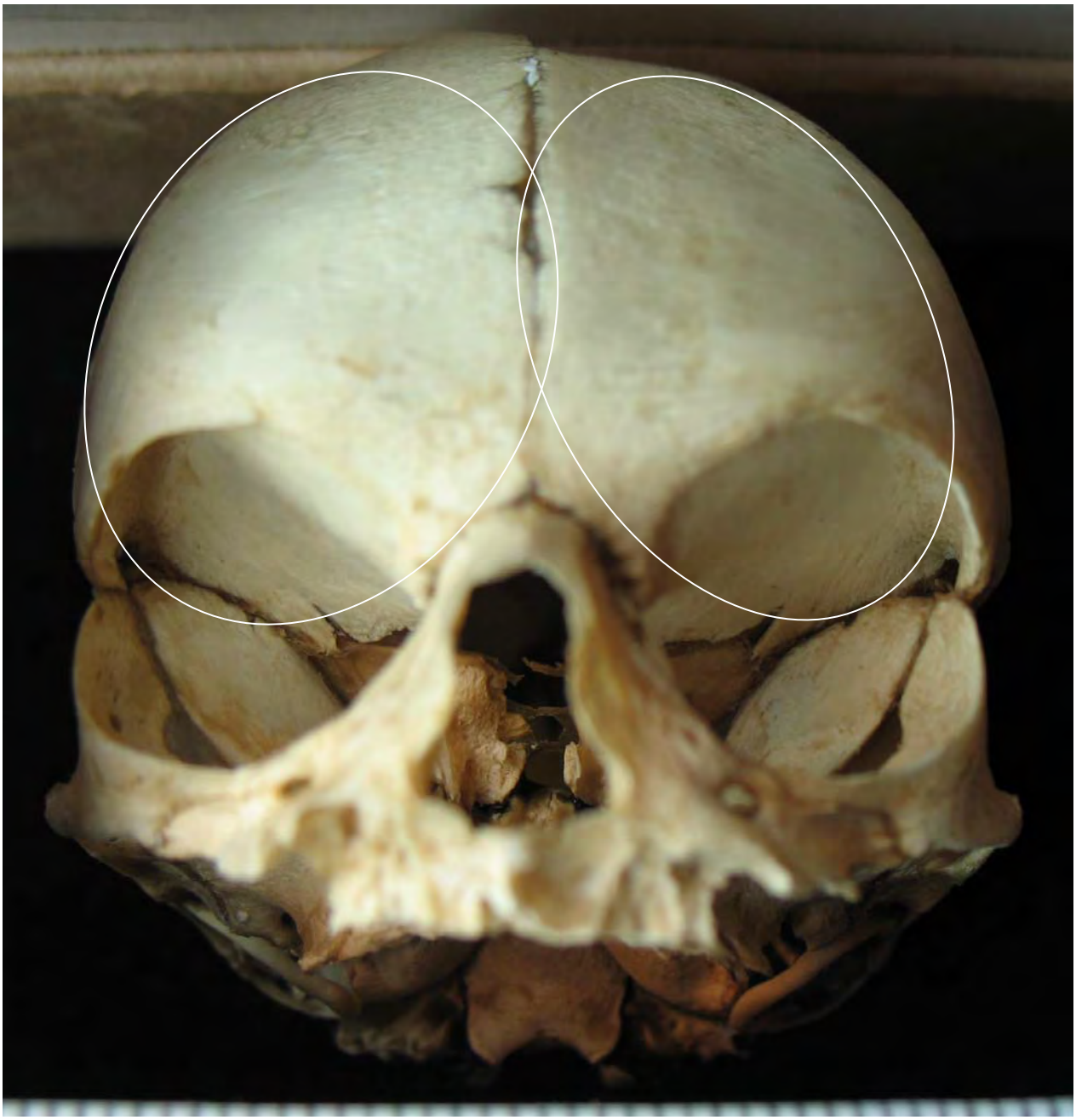


Figure 2.10: *An* articulated, but unfused, fetal cranium (23 weeks gestation). The frontals are indicated by ovals.

Sphenoid

The sphenoid is essentially the keystone bone to the skull. It fits in the center of the skull making up the posterior of the eye orbit and part of the cranial base (Figure 2.11). It articulates with the ethmoid, vomer, frontals, zygomatics, parietals, temporals, and occipital. During normal fetal growth several centers of ossification will appear, developing into the body, lesser wings, and greater wings.

Prior to the unification of the sphenoid it develops out of five parts: the central body, two lesser wings, and two greater wings (Figure 2.12). Under normal conditions the body and two lesser wings should fuse first, around 32 weeks gestation (Figure 2.13). The greater wings fuse during the first year of life to the already fused body and lesser wings.

The ossification of the sphenoid body begins during the 12th week of intra-uterine life. This bone forms endochondrally with the emergence of two primary ossification centers, one anterior and one posterior. Also during the 12th week the lesser wings begin to ossify from two centers in a cartilaginous precursor (Baker et al., 2005). Fusion of the two primary centers in the body occurs at 24 weeks creating a unified body. The greater wings of the sphenoid ossify from both endochondral and membranous tissue beginning in the 9th week (Scheuer and Black 2000, 2004). Although the greater wings begin to ossify before the body and lesser wings, they do not fuse until after birth.

Figure 2.11: Inferior view of an articulated, but unfused, fetal cranium (23 weeks gestation). The sphenoid is indicated by a rectangle.

Figure 2.12: Fetal sphenoid (32 weeks gestation) displaying the five segments: the great wings (top), the body, (middle) and the lesser wings (bottom) prior to fusion.

Figure 2.13: Fetal sphenoid (33 weeks gestation) displaying the fusion of the body to the lesser wings. Note that the greater wings have yet to fuse.

Zygomatics

The zygomatics are a robust bone that form part of the inferior eye orbit and are commonly known as the cheek bones. They articulate with the frontals, the greater wings of the sphenoid, the maxillae, and the temporal (Figure 2.14).

These bones develop from a single ossification center in the mesenchymal tissue during the 8th gestational week (Scheuer and Black 2000, 2004). Ossification spreads very rapidly in the zygomatics as it bears resemblance to the adult bone one week after beginning ossification. The bone is fully ossified and resembles its adult shape by the 4th month of intra-uterine life (Baker et al., 2005).

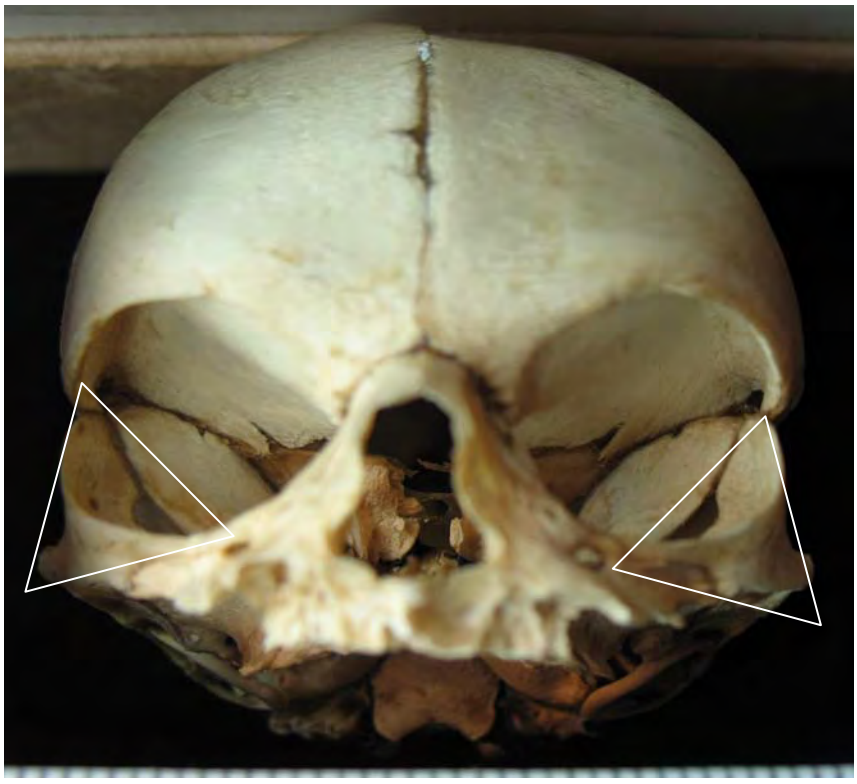


Figure 2.14: *An* articulated, but unfused, fetal cranium (23 weeks gestation). The zygomatics are indicated by triangles.

Scapulae

The scapula is part of the shoulder girdle and articulates with the clavicle and the humerus (Figure 2.15). This bone's development is described along with the previous cranial elements affected by neural tube defects because certain malformations of it are associated with craniorachischisis which extends the anencephalic defect down the fetus' spine.

The primary ossification center for the scapula appears endochondrally in the surgical neck around 7 weeks gestation (Baker et al., 2005). Spreading of osseous tissue does not occur as rapidly as it does in the cranial bones. Around the 9th week ossification has spread down the neck to its base, and in the 12th week the glenoid mass begins to ossify. The space between these two features fills in during week 14 resulting in a recognizable structure that resembles the adult bone (Scheuer and Black, 2000, 2004). The epiphyses for the scapula do not begin to ossify until after birth.

Figure 2.15: Posterior view of fetal scapulae (32 weeks gestation).

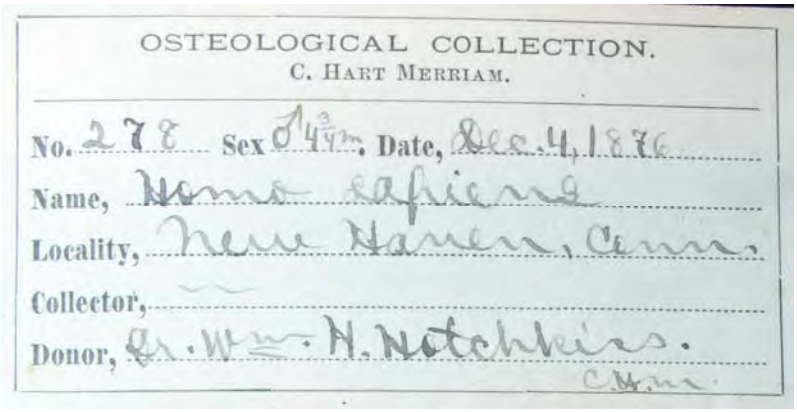
Materials and Methods

Twenty six non-pathological fetuses were randomly selected and analyzed along with nine documented anencephalic skeletons housed at the NMNH Washington, DC for the purpose of comparing clinical literature with bony elements that could possibly be found archaeologically

Table 2.1: Individuals included in NMNH sample. Information included is all that is provided to researchers about the individuals in the collection.

CAT#	Collector	Sex	Age	Ethnicity	Notes Provided
218141	Lamb, DS	Male	06	White	About 6.5 Months, Anencephaly, Craniorachischisis, Spina Bifida
218142	Lamb, DS	Female	07	White	About 7 Months, Pseudencephalus, Spina Bifida
218143	Lamb, DS	Male	09	White	At term, Anencephaly
218144	Lamb, DS	Female	06	Mixed	About 6 months
218146	Lamb, DS	Unknown		White	Anencephaly
219199	Lamb, DS	Female		White	About 7 Months, Anencephaly, Spina Bifida, Craniorachischisis
219200	Lamb, DS	Male	09	White	Apparently at term, Anencephaly
220182	Lamb, DS	Female		White	
222089	Lamb, DS	Female		White	Twins with 222090
224710	Scurlock, HC	Male		Black	Brain Saved (224709)
224741	Morgan, EL	Male		White	
224855	Mall, FP	Male		Mixed	Premature, About 8 months, Died 3 days after birth
224859	Mall, FP	Female		Mixed	Full term, New Born, Died 6 days after birth
224884	Mall, FP	Male		Mixed	Anencephalic
224885	Mall, FP	Female		Mixed	
224888	Morgan, EL	Female	07	Mixed	About 7-8 months
226550	Hrdlička, A	Female		White	
226553	Hrdlička, A	Unknown		Unknown	Anencephalic
228369	Poole, BG	Male	06	White	About 6-7 months, Brain Saved (228368)
228839	Mall, FP	Female		White	
249557	Mall, FP	Male	09	White	New Born
250421	Buchanan, WR	Male	04	White	4.5 Months, Brain Saved (250422)
253843	Lane, TF	Male	04	White	4 Months Premature Birth
253850	Lane, TF	Female		White	
253851	Lane, TF	Male		White	
253858	Lane, TF	Female	03	Black	3 Months
271013	n/a	Unknown		Unknown	
290932	Smith, JH	Male		White	Anencephalic
299234	Smith, JH	Female		White	
299308	Holmes, JH	Male		White	
299312	Holmes, JH	Male		White	
299443	Holmes, JH	Female		White	
299453	Holmes, JH	Male		Black	
381034	Holmes, JH	Unknown	04	Unknown	4.5 Months
T007	n/a	Unknown		Unknown	

(Table 2.1). Most of these remains were collected by pathologists, embryologists, and private physicians during the late 19th and early 20th century from lower socioeconomic groups in the Balitmore/DC area, and curated by Ales Hrdlička between 1903 and 1917 (Gindhart, 1989). Currently the collection at the NMNH consists of nearly 400 individuals, aged from 7 weeks gestation through 10 months after birth. A bias is represented in anomalous and/or pathologic remains based on the fact that the majority of the collection resulted from spontaneous abortion (Huxley, 2005).


OSTEOLOGICAL COLLECTION.
C. HART MERRIAM.

No. 278 Sex ♂ 4m. Date, Dec. 4, 1876

Name, Homo sapiens

Locality, New Haven, Conn.

Collector,

Donor, Dr. Wm. H. Hotchkiss.

Figure 2.16: Example of the "medical records" provided to researchers.

Unfortunately, no medical records (Figure 2.16) exist regarding the individuals or their mothers, and listed age estimations at the time were based on cephalic circumference, in conjunction with torso and appendage length. Ales Hrdlička, the curator at the time, noted the information available, which included sex, estimated age, and some randomized external measurements (Gindhart, 1989). At the time of analysis, the NMNH had 6, possibly 7, anencephalic skeletons available for examination. Through the course of analysis it was determined that the 7th questionable skeleton did in fact display classic characteristics of anencephaly. Two additional anencephalic skeletons were discovered in the collection, bringing the total sample size to 9.

Altogether 9 anencephalic and 26 non-pathological fetuses from the NMNH were analyzed. Of the NMNH sample, 16 were male, 14 were female, and the sex of 5 are unknown (Figure 2.17). Twenty-two of these were listed as white, 3 as black, 6 as colored, mixed or mulatto, and the ethnicity of 4 are unknown (Figure 2.18). Digital calipers were used to take 12

cranial and post-cranial measurements (Table 2.2). Measurements were recorded to the nearest hundredth of a millimeter in order to assess age. These measurements were then compared to the data published by Scheuer and Black (2000, 2004) to provide an age estimate (Table 2.3). Sherwood and colleagues (2000) demonstrated that the measurements taken from fetuses with anencephaly caused them to over age the individuals by 1 to 2 weeks. Their estimate was offset by the average difference of two weeks between gestational age (or weeks since the mother's last normal menstrual period) and time since conception. Based on their research it is possible that the estimation of age in the anencephalic individuals from the NMNH in this study could be underestimated by one to two weeks.

Table 2.2: Measurements taken from all individuals included in NMNH sample. All measurements taken were from digital calipers and are based on descriptions provided by Scheuer and Black (2000).

Bone	Measurement	Description
Basion	Length	Midsagittal distance from anterior border of foramen magnum to anterior border
Basion	Width	Maximum width at level of lateral angles
Zygomatics	Width	Medial end of infra-orbital border to superior end of frontal process
Clavicle	Length	Maximum length of the entire bone
Scapula	Height	Distance between the superior and inferior angles
Scapula	Width	Distance between margin of glenoid fossa and medial end of spine
Humerus	Length	Maximum length of the entire bone
Ulna	Length	Maximum length of the entire bone
Radius	Length	Maximum length of the entire bone
Femur	Length	Maximum length of the entire bone
Tibia	Length	Maximum length of the entire bone
Fibula	Length	Maximum length of the entire bone

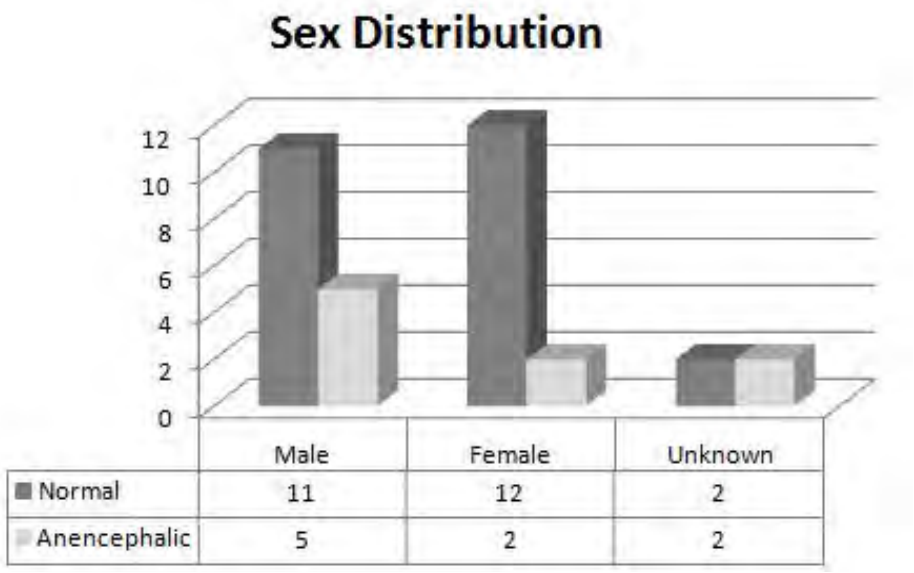


Figure 2.17: The total sex distribution of the NMNH sample.

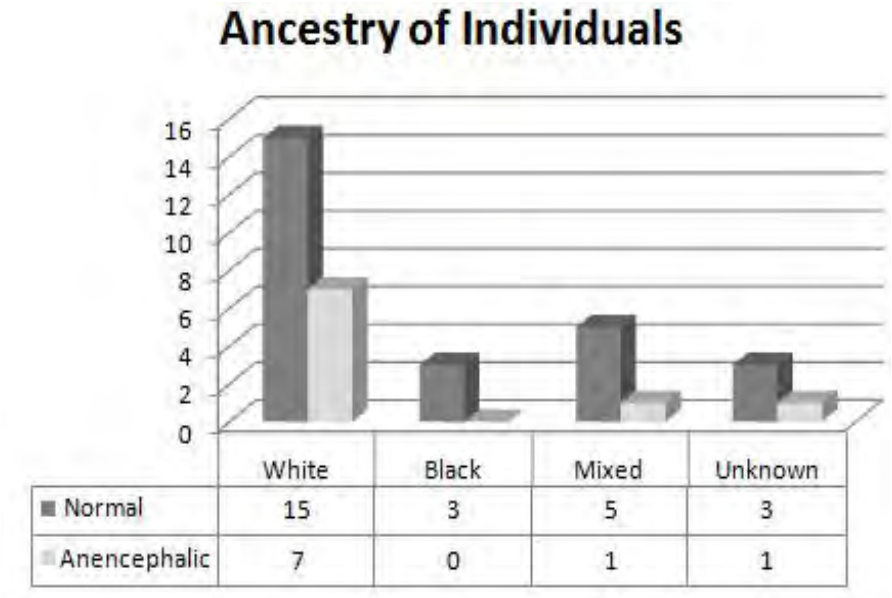


Figure 2.18: Ancestry of the total NMNH sample.

Table 2.3: This table shows the beginnings of ossification (X), and the gestational week in which the fetal bone resembles the adult morphology (O).

Developing Fetal Bones Morphologically Effected by Anencephaly	Gestational Weeks																																						
	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31	32	33	34	35	36	37	38	39	40
PARIETALS							X													O																				
OCCIPITAL																																								
Squamous																																							
pars interparietalis								X												O																				
pars supra-occipitalis									X											O																				
Pars Lateralis								X																																
Pars Basilaris											X																													
TEMPORALS																																		O						
Petrous																X																								
Squamous							X																															O		
Tympanic Ring									X																													O		
FRONTALS													O																											
Squamous						X																																		
Supercilliary arch										X																														
Zygomatic processes										X																														
Orbital plate								X					O																											
SPHENOID																																								
Body												X												O																
Lesser wings												X																				O								
Greater wings									X																															
Zygomatics								X									O																							
Scapulae							X							O																										

Results and Discussion

The nine anencephalics at the NMNH were measured as to asses age (Figure 2.19) in order to be compared to K2 B563 and a review of clinical literature has provided the basis for the necessary qualitative analysis of the anencephalic individuals from the NMNH collection (Table 2.4).

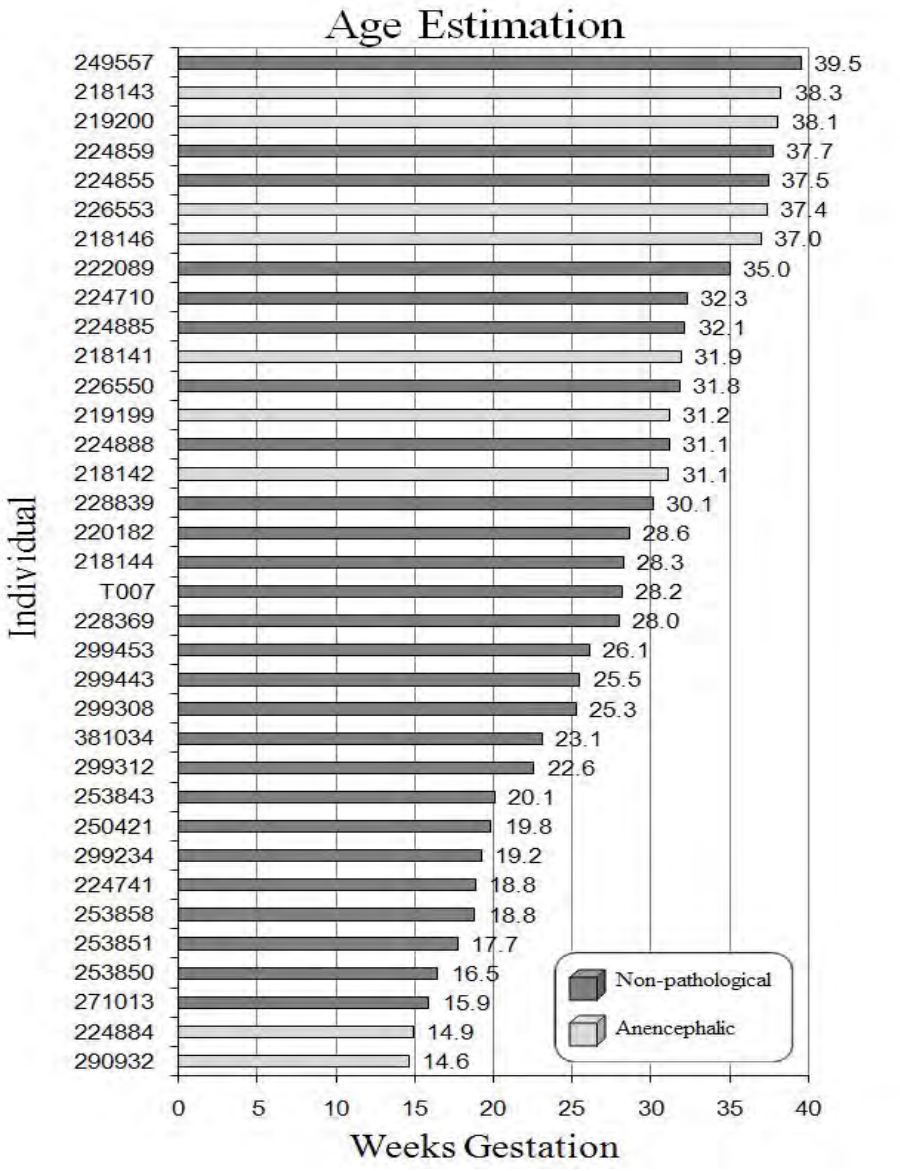


Figure 2.19: Estimated age in weeks gestation for the total sample from the NMNH.

Table 2.4: Summary of comparison of cranial bones showing the difference in morphology between normal development and anencephaly.

Cranial Bones	Non-pathological	Anencephalic
PARIETALS	2 unfused elements. Elliptical in shape and convex exteriorly with serrated borders.	Complete absence of bone.
OCCIPITAL	4 unfused elements.	Variable number of elements.
Squamous	Shallow bowl shaped appearance with a prominent external occipital protuberance at the junction.	If any bone from squamous is present it is unfused and uncharacteristic in shape.
pars interparietalis	Fan shaped with thin feathery borders.	Complete absence of bone.
pars supra-occipitalis	Wedge shaped with thick serrated borders.	If this bone does ossify it is undersized. Robust. Appears as a small wedge or sickle shape with a feathered process extending medially and superiorly.
Pars Lateralis	A thickened medial border and a wedge shaped posterior border. 2 inferior limbs extends medially to create the hypoglossal canal.	Fairly similar in shape to normal fetuses, although the width of the bone is more robust and either disproportionately wide or narrow.
Pars Basilaris	Robust and square with a D-shaped cut out at the superior surface. The inferior surface is slightly concave. Wider than it is long.	Similar in shape to normal fetuses although the proportions are inverted: the bone is longer than it is wide.
TEMPORALS	2 unfused elements on either side.	Early fusion of all elements.
Petrous	Cylindrical in shape. Medially the internal acoustic meatus is anterior to the large subarcuate fossa. Superiorly the surface is smooth.	Highly irregular in shape and shorter in total length than normal. Foramen appear smaller and are located irregularly.
Squamous	A delicate, nearly flat, semicircular plate with serrated borders. The zygomatic process extends anteriorly.	Appears to have curled back upon itself onto the petrous portion. The zygomatic process is the only portion that is recognizable, yet is shortened.
Tympanic Ring	A nearly complete circle of bone in the shape of a ring fused to the squamous.	Fuses early. Appears more elliptical than circular.
FRONTALS	2 unfused elements. An irregular, yet bowl shaped bone. The superior squamous portion is rounded and convex exteriorly. The orbital plate meets the squamous inferiorly with a dome shape.	Complete absence of the squamous portion. Only the anterior border of the orbit forms, with an excessive curvature. The shape of this bone is the most variable in anencephalics.
SPHENOID	3 unfused elements.	Early fusion of all elements.
Body	Cube shaped with 2 alar processes which resemble feathers.	Far more narrow than normal. Almost rounded. Usually longer and far narrower than usual.
Lesser wings	Flat and arrow-head shaped bone which fuses to the body.	Nearly unrecognizable. Curled back upon the body. Severely undersized.
Greater wings	Triangular in shape this bone's lateral $^{2}/_{3}$ are thin and slightly concave anteriorly. The medial $^{1}/_{3}$ is a complex root for attaching to the body.	Fuses to the body early. The malformations are nearly indescribable. The root is wider and more robust whereas the wing is also undersized and shows the "curled" effect.
ZYGOMATICS	A robust bone resembling an upside down letter T. The superior process extends towards the frontals, the anterior process extends towards the maxilla, and the posterior process extends toward the temporal.	Appears normal with the exception of the complete or nearly complete absence of the temporal process. Much more robust than normal.

During data collection specific anatomic anomalies associated with anencephaly were noticed. This study revealed that all of the 9 anencephalic fetuses were missing both parietals. The anencephalics were also missing the squamous portion of the occipital, or only a minor fragment of the supra-occipitalis was present. Furthermore, the anencephalic fetuses' pars lateralis and pars basilaris were malformed. The shape of this anomaly varied between the 9 individuals analyzed (Figure 2.20).

Figure 2.20: Disarticulated anencephalic fetal occipitals (32 weeks gestation) comparing normal development (left) to the development of an anencephalic (right). On the anencephalic, note the absence of the pars interparietalis, and the hindered growth of the pars supra-occipitalis. Also note the abnormal shape of the basion.

In the NMNH collection the anencephalic frontal bones only exhibit the portion that makes up the superior portion of the eye orbit, and are completely missing the squamous portion (Figure 2.21).

Figure 2.21: Fetal frontals (39 weeks gestation) comparing normal development (left) to the development of an anencephalic (right). On the anencephalic, note the absence of the squamous portion and the excessive curvature of the roof of the eye orbit.

As discussed previously, during normal development, the temporals are comprised of three elements which fuse to form a single bone shortly after birth. The data shows that the temporals of these anencephalic individuals fused prematurely, sometime after 14 weeks and before 29 weeks gestation. Even though the three elements fuse early, the squamous portion of the temporal is nearly missing; the only feature that remains is a shortened zygomatic process (Figure 2.22).

Figure 2.22: (Right side) Articulated fetal temporal (39 weeks gestation) comparing normal development (left) to the development of an anencephalic (right). On the anencephalic, note the absence of the squamous portion and the hindered growth of the petrous portion.

The temporal processes of the anencephalic zygomatics were either missing altogether, or were very small and broad (Figure 2.23).

Figure 2.23: (Right side) Fetal zygomatic (32 weeks gestation) comparing normal development (left) to the development of an anencephalic (right). On the anencephalic, note the absence of the temporal process.

The most deformed of the anencephalic bones is the sphenoid. As discussed previously, this bone is comprised of a body, two lesser wings, and two greater wings. During normal development, the lesser wings fuse to the body during the 20th week of gestation, and the greater wings fuse to the body during the first year of life. All of the NMNH anencephalic fetuses' lesser wings fused prematurely to the body and were so malformed that they were nearly unrecognizable. Most of these fetal skeletons' greater wings had also fused prematurely, and all of the greater wings are severely misshapen (Figure 2.24).

Figure 2.24: Fetal sphenoid (33 weeks gestation) comparing normal development (left) to the development of an anencephalic (right). On the anencephalic, note the early fusion of the greater wings and that both the greater and lesser wings are severely misshapen.

Three of the nine anencephalic skeletons also had the defect craniorachischisis (218141, 218142, and 219199), which distorted the measurements of their clavicles and scapulae (Figure 2.25). Yet, the measurements of all the long bones appear to represent normal fetal growth. These measurements are comparable to the other fetal skeletons analyzed from the collection at the NMNH.

Figure 2.25: Scapulae (28 weeks gestation) of a normal fetus (left) and from an anencephalic fetus with craniorachischisis (right). Note that the superior border is missing on the right.

While analyzing the craniofacial elements of the anencephalics at the NMNH it was apparent that this NTD distorted the morphology of the bones. Therefore making age estimation based solely on a comparative assessment of previous studies of fetal cranial growth and the present individuals would be impossible. Figure 2.26 displays they widely varying age estimates, in weeks gestation, for each individual analyzed both anencephalic (left) and non-pathological (right). Here you can see, for example, that anencephalic individual 218142's length of the basion suggests 38 weeks gestation, whereas the width of the same bone suggests an age of 25 weeks gestation. Although several anencephalic individuals vary drastically in age estimation based craniofacial skeletal measurements, not all individuals from this sample display such a drastic variation in age as individual 218142. It is being suggested that this is caused by a range in severity in anencephaly. Therefore, the more severe the defect the more drastically the measurements used to estimate age are varied.

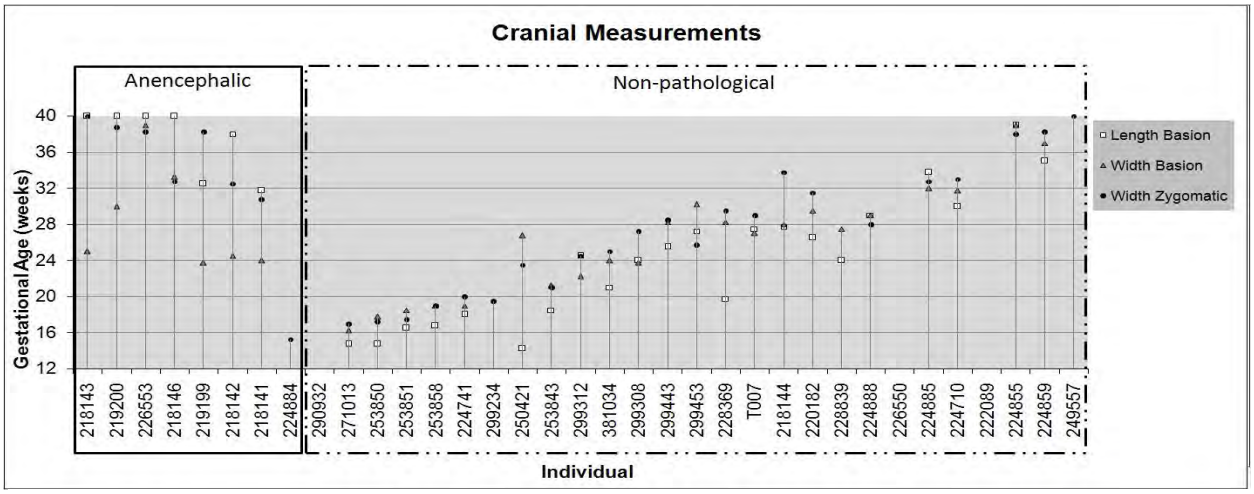


Figure 2.26: Comparing the cranial measurements of anencephalic fetuses (left) to non-pathological fetuses (right).

When the age estimates based on cranial measurements are directly compared among the non-pathological specimens very few individuals displayed this drastic range of variation. It is possible that the variation seen in these few individuals could suggest some unknown pathology that causes bones malformation, or it could simply be an error in measurement. Generally, the analysis of the cranial age estimations alongside the appendicular age estimations (Figure 2.27) shows little variation in age in both the nonpathological individuals and the anencephalics, with the exception of those with craniorachischisis (226553A, 219199, and 218141). These findings further support that the postcranial skeleton develops normally in individuals affected by the NTD anencephaly.

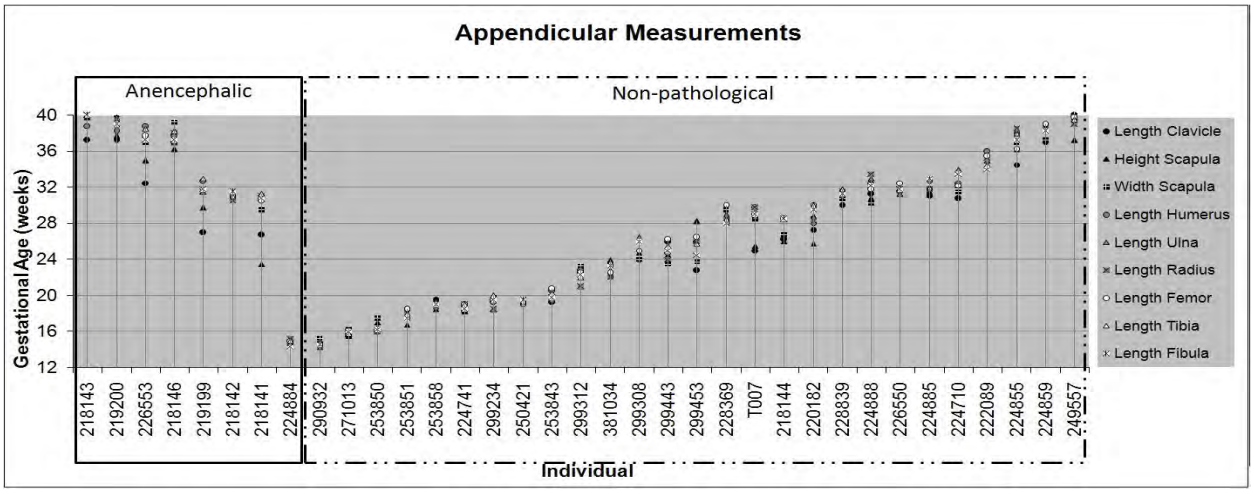


Figure 2.27: Comparing the appendicular measurements of anencephalic fetuses (left) to non-pathological fetuses (right).

Conclusion

Neural tube defects are developmental errors that occur very early in pregnancy. Although some NTDs are ultimately fatal, others can be surgically corrected while still in the mother's uterus. Modern legislation has mandated the fortification on grain products with the B-vitamin folate with the intention of preventing further occurrences of NTDs. Further research would be advantageous in the clinical analysis of modern anencephalics' skeletons as the NMNH collection is based on lower socioeconomic groups from nearly one hundred years ago.

The compilation of qualitative descriptors from the NMNH collection based on the comparative analysis of each cranial element affected by anencephaly in both non-pathological and anencephalic individuals helped to establish criteria for a diagnostic tool for the diagnosis of the skeletal characteristics of anencephaly. It is possible for individuals with these characteristics to be diagnosed from an archaeological context. Should a fetal skeleton be excavated without parietals, the squamous portion of the frontals, temporals, or occipital, and a nearly unrecognizable prematurely fused sphenoid a bioarchaeologists could assess the possibility of anencephaly.

References

Abd-El-Malek S. 1957. The anencephalic skull and a vascular theory of causation. *Egyptian Medical Association.* 40:216-224.

Baker BJ, Dupras TL, and Tocheri MW. 2005. *The Osteology of Infants and Children*. College Station: Texas A&M University Press.

Bailey LB, Rampersaud GC and Kauwell GPA. 2003. Folic Acid Supplements and Fortification Affect the Risk for Neural Tube Defects, Vascular Disease and Cancer: Evolving Science. *The Journal of Nutrition*. 133: 1961S-1968S.

Barnes E. 1994. *Developmental Defects of the Axial Skeleton in Paleopathology*. Niwot: University Press of Colorado.

Charon P. 2005. Teratology of the neural tube: history and paleopathology. *Antropo.* 10:83-101.

Daly S. and Mills JL. 1997. Minimum Effective Dose of Folic Acid for Food Fortification to Prevent Neural-Tube Defects. *Lancet*. 350(9092):1666-1669.

Dambska M. and Wisniewski KE. 1999. *Normal and Pathologic Development of the Human Brain and Spinal Cord*. London: John Libbey & Company Ltd.

Dolk H., De Wals P., Gillerot Y., Lechat MF., Ayme S., Cornel M. and Cuschieri A. 1991. Heterogeneity of Neural Tube Defects in Europe: The Significance of Site Defect and Presence of Other Major Anomalies in Relation to Geographic Differences in Prevalence. *Teratology*. 44:547-559.

Erickson JD. 2002. Folic Acid and Prevention of Spina Bifida and Anencephaly: 10 Years After the U.S. Public Health Service Recommendation. Morbidity and Mortality Weekly Report. 51: 3-5.

Fields HW., Metzner L., Garol JD., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses I. Cranial Floor. *Teratology*. 17:57-66.

Garol JD., Fields HW., Metzner L., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses II. Calvarium. *Teratology*. 17:67-74.

Geoffroy Saint-Hillaire I. 1847. Vie, Travaux et Doctrine Scientifique d' Étienne Geoffroy Saint-Hillaire par son Fils. Paris: Bertrand.

Giroud A. 1960. Causes and morphogenesis of anencephaly. In: *Ciba Foundation Symposium on Congenital Malformations*. GEW Wolstenholme and M O'Connor, eds. J and A Churchill, London. 199-218.

Green NDE. and Copp AJ. 2006. The Embryonic Basis of Neural Tube Defects. In: Diego Wyszynski, editor. *Neural Tube Defects*. New York: Oxford University Press p15-28.

Gindhart PS. 1989. An early twentieth-century skeleton collection. *Journal of Forensic Sciences*. 34(4):887-893.

Hibbard ED. and Smithells RW. 1968. Folic Acid Metabolism and Human Embryopathy. *Lancet*. 1254.

Huxley AK. 2005. Gestational Age Discrepancies Due to Acquisition Artifact in the Forensic Fetal Osteology Collection at the National Museum of Natural History, Smithsonian Institution, USA. *American Journal of Forensic Medicine & Pathology*. 26(3):216-220

Isada NB, Qureshi F, Jacques SM, Holzgreve W, Tout MJ, Johnson MP, and Evans MI. 1993. Meroanencephaly: pathology and prenatal diagnosis. *Fetal diagnosis and therapy*. 8(6):423-428.

Keen JA. 1962. Morphology of the skull in anencephalic monsters. *South African journal of Laboratory and Clinical Medicine.* 8:1-9.

Langman J, and Welch GW. 1966. Effect of Vitamin A on the development of the central nervous system. *Journal of comparative Neurology.* 128:1-16.

Lemire RJ. 1988. Neural Tube Defects. *Journal of American Medical Association*. 259:558-562.

Lewis ME. 2007. *The Bioarchaeology of Children*. Cambridge: Cambridge University Press.

Larsen WJ. 2001. *Human Embryology*. Philadelphia: Churchill Livingstone.

Lie RT. 2006. An International Perspective on Anencephaly and Spina Bifida, Prevalences by the Turn of the Century. In, Wyszynski DF (ed.) *Neural Tube Defects*. New York: Oxford University Press.

Marin-Padilla M. 1965. Study of the skull in human cranioschisis. *Atca Anatomy.* 62:1-20.

Mathews TJ., Honein MA. and Erickson JD. 2002. Folic Acid and Prevention of Spina Bifida and Anencephaly: 10 Years After the U.S. Public Health Service Recommendation. Morbidity and Mortality Weekly Report. 51: 11-13.

Merker B. 2007. Consciousness without a cerebral cortex: A challenge for neuroscience and medicine. *Behavioral and Brain Sciences.*30:63-81.

Metzner L., Fields HW., Garol JD., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses III. Facial Skeleton. *Teratology*. 17:75-82.

Mills JL. and England L. 2001. Food Fortification to Prevent Neural Tube Defects: Is It Working? *The Journal of the American Medical Association*. 285(23): 1172-1173.

Moore CA. 2006. Classification of Neural Tube Defects. In, *Neural Tube Defects*. Wyszynski DF; editor. New York: Oxford University Press.

Moore Ca, Li S, Hong S, Gu H, Berry RJ, Mulinare, J, and Erickson JD. 1997. Elevated rates of severe neural rube defects in a high-prevalence area in Northern China. *American Journal of Medical Genetics.*73(2)113-118.

Naggan L, and MacMahon B. 1967. Ethnic differences in the prevalence of anencephaly and spina bifida in Boston, Massachusetts. *New England Journal of Medicine*. 227:1119-1123.

Parker Pearson M. 1999. *The Archaeology of Death and Burial*. College Station: Texas A&M University Press.

Quinlivan EP and Gregory JF. 2003. Effect of Food Fortification on Folic Acid Intake in the United States. *American Journal of Clinical Nutrition*. 77: 221-225.

RobertsC and Manchester K.1995. *the Archaeology of Disease*. Ithaca: Cornell University Press.

Sadler TW. 2006. *Medical Embryology*. Philadelphia: Lippincott Williams and Wilkins.

Scheuer L, and Black S. 2004. *The Juvenile Skeleton*. London: Academic Press.

Scheuer L. and Black S. 2000. *Developmental Juvenile Osteology*. London: Academic Press.

Shaffer LG, Marazita ML, Bodurtha J, Newlin A, and Nance WE. 1990. Evidence for a major gene in familial anencephaly. *American Journal of Medical Genetics.*36:97-101.

Shane B. 2003. Folate Fortification: Enough Already? *American Journal of Clinical Nutrition*. 77: 8-9.

Sherwood RJ., Meindl RS., Robinson HB., and May RL. 2000. Fetal Age: Methods of Estimation and Effects of Pathology. *American Journal of Physical Anthropology.* 113:305-315.

Stevenson AC, Johnston HA, Stewart MIP, and Golding DR. 1966. Malformation of structures developed from the neural tube. *Bulletin of the World Health Organization*. 34(Suppl.9):25-34.

Ulrich CM. and Potter JD. 2006. Folate Supplementation: Too Much of a Good Thing? *Cancer Epidemiology Biomarkers and Prevention*. 15(2): 189-193.

Walsh DS. and Adzick NS. 2003. Foetal Surgery for Spina Bifida. *Seminar in Neonatology*. 8(3):297-305.

Wharton B and Booth I. 2001. Fortification of Flour with Folic Acid: A Controlled Field Trial is Needed. *British Medical Journal*. 323:1198-1199.

Williams LJ., Mai CT., Edmonds LD, Shaw GM., Kirby RS., Hobbs CA., Sever LE., Miller LA., Meaney FJ., and Levitt M. 2002. Prevalence of Spina Bifida and Anencephaly During the Transition to Mandatory Folic Acid Fortification in the United States. *Teratology*. 66: 33-39.

CHAPTER 3
ANENCEPHALY IN THE KELLIS 2 CEMETERY IN DAKHLEH OASIS, EGYPT

Introduction

The existence of human fetal skeletal remains in the archaeological record is unusual. The discovery of well preserved fetal remains displaying skeletal birth defects are an even rarer occurrence. The fragility of fetal remains makes them highly vulnerable and they are most often destroyed as a part of the decomposition process or careless excavation technique. Human fetal remains are also uncommon in modern osteological collections because they are frequently misidentified and labeled as non-human. This misidentification is particularly common in cases with fragmentary remains and in areas with poor preservation (Scheuer and Black, 2000). In rare cases the environment and past cultural practices may aid in the preservation of fetal remains. The arid climate, high alkalinity of the soils, and cultural practices of the ancient peoples in the Dakhleh Oasis, Egypt, have promoted incredible preservation of human remains including a large collection of fetal remains. One of these fetal individuals, K2 B563, appears to have the classic anencephalic skeletal features.

The Dakhleh Oasis is one of the largest of five major oases in Egypt's western desert and is located some 725km SSW of Cairo (Dupras and Tocheri, 2007). Interdisciplinary research in the Dakhleh Oasis is conducted under the Dakhleh Oasis Project (DOP), whose core mandate is

to better understand the biocultural interaction of the ancient people of Dakhleh and the harsh desert environment (Mills, 1984). Currently the majority of the bioarchaeological research takes place in one of two cemeteries associated with the ancient town of Kellis. This cemetery is known as the East or Kellis 2 cemetery. Archaeological data suggests that the town of Kellis was occupied during the late Ptolemaic and Roman periods (c. 31BC - 360AD) (Hope, 2001).

The Kellis 2 cemetery contains a large sample of remarkably preserved skeletons interred at varying depths, which seem to correspond with the individuals' height (Tocheri et al., 2005). Almost every burial follows a recurring pattern: a separate grave for each individual with very few, if any, grave goods, laid to rest in a supine position oriented with head facing west (Birrell, 1999). Five radiocarbon dates from skeletal material within Kellis 2 indicate that the cemetery was in use between AD 100 and 450 (Fairgrieve and Molto, 2000; Stewart et al., 2003). Note that there is a discrepancy between the dating of Kellis and the Kellis 2 cemetery – this is currently being debated in the DOP (pers comm., Dupras). This combined evidence appears to indicate an early Christian style of burial (Bowen, 2003) and is mirrored in other early Christian sites (Parsche and Zimmerman, 1991).) Furthermore, the presence of fetal remains buried alongside other individuals in the cemetery is documented in early Christian burials (Marlow, 2001). It is hypothesized that the spatial distribution of individual graves within the cemetery is correlated to family plots (Molto, 2001; Tocheri et al., 2005).

In 2003 the remains of a fetal individual (B563) were excavated from the southeastern sector of the Kellis 2 cemetery. Upon analysis this individual displayed very specific cranial pathologic defects that appear to be related to a neural tube defect. The purpose of this research

is to analyze the human fetal skeleton K2 B563 from the Kellis 2 cemetery in order to ascertain whether it displays the classic characteristics of anencephaly. It is imperative to rule out 'breakage' or 'decomposition' as the reason for this type of skeletal defect (Miller and Simon, 2001). To make this determination, the skeleton from the Dakhleh Oasis was compared to a collection of nine fetal skeletons with clinically diagnosed anencephaly, housed at the Smithsonian Institution's National Museum of Natural History (NMNH), in Washington D.C.

Background of Neural Tube Defects

The focus of this research is whether an archaeological specimen, K2 B563 from the Dakhleh Oasis, Egypt, displays classic skeletal characteristics of the neural tube defect anencephaly. To diagnose this condition in an unknown archaeological skeleton, a clinical comparison is needed. To date, the Hermopolis mummy, so named for being found in Hermopolis Egypt, is the only published archaeological example of an anencephalic fetus, and it was last described in 1847 (Saint-Hillaire, 1847). Although there is an abundance of published clinical descriptions of the soft tissue formation associated with anencephaly, there is little that describes the skeletal characteristics. Due to a lack of published sources documenting the skeletal diagnosis of anencephaly it is necessary to draw conclusions based on modern clinical skeletons that display this condition.

Understanding embryology, in terms of bone growth patterns, is a crucial skill necessary for determining how and when developmental errors occur. Neural tube (NT) defects originate

in the third and fourth week after conception as a result of the failure of neurulation, or proper closure of the neural tube at the cranial or caudal end. This is the definitive point during development when a number of severe cranial or spinal defects often occur, and sometimes both. Malformations caused by the failure of the neural tube to close properly include NTDs such as spina bifida and anencephaly. Neural tube defects are fairly common developmental abnormalities of the central nervous system that can be life threatening (Green and Copp, 2006).

The cause of neural tube defects remains speculative. Research on the origins of NTDs is ongoing considering both genetic and environmental factors, with supporting evidence for both (Shaffer et al., 1990). Although the medical community recognizes the various types of NTDs, variation within each type is poorly understood (Moore, 2006). Spina bifida and anencephaly are among the most common of NTDs. Because detailed data regarding the form or type of NTD present at birth is generally not recorded, little is known about the potential demographic differences between the types. Both untreated severe forms of spina bifida and anencephaly usually result in a premature birth. If the fetus survives to term, most individuals do not live beyond a few hours or 2-3 days at most (Dambska and Wisniewski, 1999).

Spina Bifida

Spina bifida is a series of neural tube defects in which the caudal neuropore remains open. This NTD is classified by the vertebral arches to failure to fuse, exposing the spinal cord (Barnes, 1994). These malformations range widely in severity. Individuals with spina bifida occulta, the mildest form, may lead normal lives without ever knowing they have a spinal defect.

Those with spina bifida cystica develop the same skeletal characteristics as those with spina bifida occulta, only the exposed spinal cord is pushed outside of the body during fetal development and is contained in a sac filled with cerebrospinal fluid. If untreated this defect will result in fatality; however, the advancement of medical technology has developed a procedure call hysterotomy in which spina bifida cystica can be corrected while the fetus is still in its mother's uterus. Meningocele is so similar in appearance to spina bifida cystica, that they are often misdiagnosed. Also known as myelschisis and myelocele, this malformation's abnormality lies with the meninges, or the membranes that envelop and protect the nervous system. In this severe and very rare type of spina bifida, the meninges are pushed out through the unfused vertebrae into a sac filled with cerebrospinal fluid, but the spinal cord develops normally and remains intact (Moore 2006).

Anencephaly

Anencephaly is a series of neural tube defects in which the cranial neuropore remains open. The clinical description of anencephaly includes a lack of cranial vault bones, which results in an exposed dorsal mass of undifferentiated neural tissue (Dambska and Wisniewski, 1999). The little descriptive research published on anencephaly tends to focus on the development of the brain, or lack thereof. There are no clinical publications specifically describing the development of the malformed cranial bones.

Anencephaly is clinically classified into three categories based on severity: meroanencephaly, holoanencephaly, and craniorachischisis. All forms of anencephaly are

equally fatal regardless of severity. Meroanencephaly is “classic” form of anencephaly and is the more common than the other two. These individuals lack portions of the cranial vault bones which exposes the brain, or lack thereof.

The defect in holoanencephalics is more severe than in meroanencephalics. In this type of anencephaly the bony defect extends from the vault through to the foramen magnum exposing the interior of the skull from the superior as well as the posterior. Craniorachischisis, the third type of anencephaly, is so rare that it is sometimes classified as a distinct defect. Fetuses with craniorachischisis display holoanencephaly extending into continuous spina bifida down to the thoracic vertebrae.

Only one example of ancient anencephaly, a mummified male fetus from Hermopolis, Egypt, has been published. This individual, excavated by archaeologist Joseph Passalacqua, was found buried amongst Old Kingdom sacred mummified baboon remains, and was originally thought to be a baboon itself. The mummy was deemed human by the French biologist who coined the term anencephaly, Étienne Geoffroy Saint-Hillaire, after being studied in detail throughout the 1820s. His publication in 1822 titled *Philosophie Anatomique des Monstruosits Humaines* devoted 50 pages of text to the description of anencephaly in several cases. Also included was a sketch of the cranial elements of an anencephalic fetus after autopsy (Figure 3.1) which Saint-Hillaire used in comparison to analyze the Hermopolis mummy. The translated description of these malformed bones was published by Pierre Charon (2002). Collins Cook

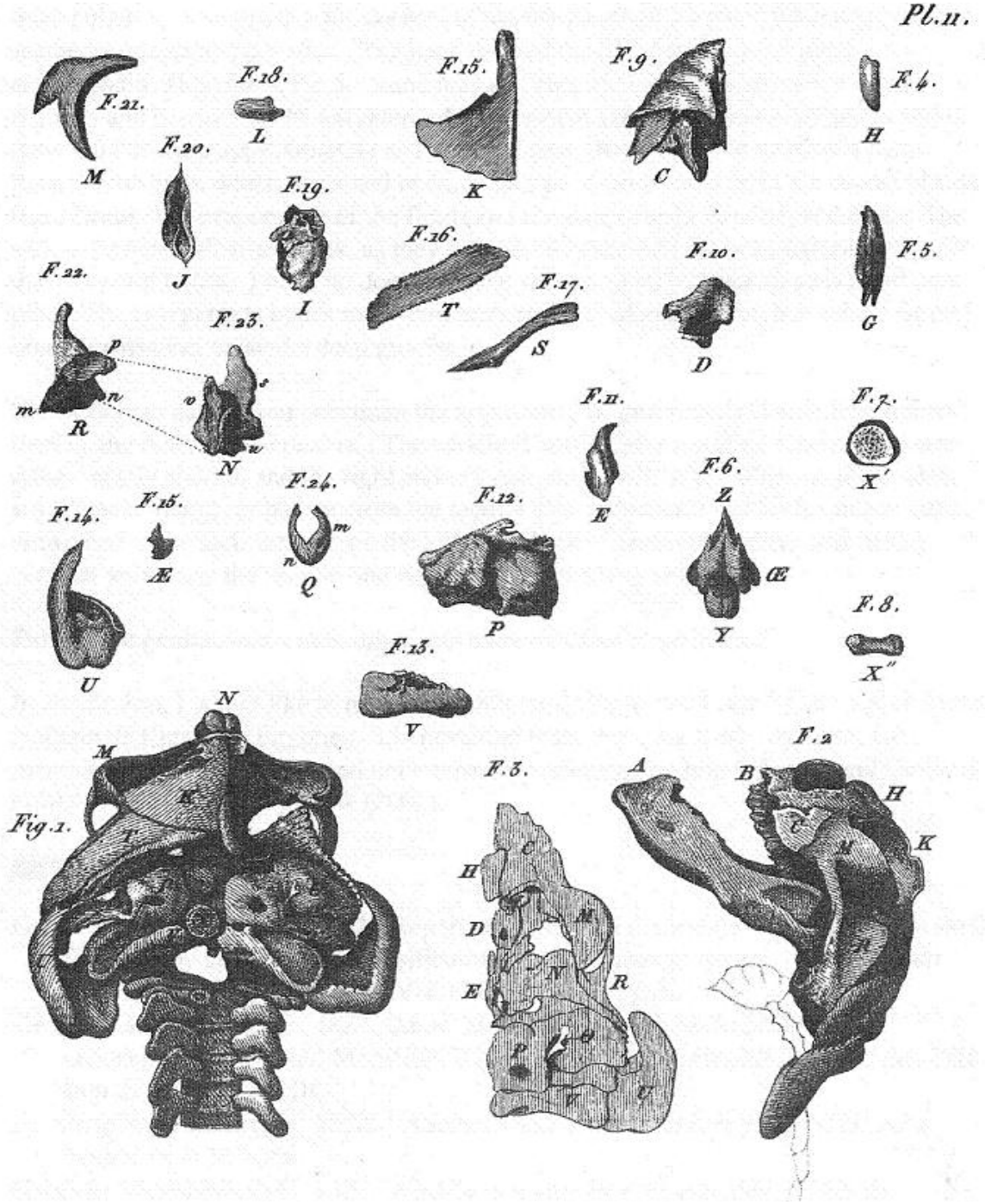


Figure 3.1: Saint-Haillarie's sketch of the cranial elements of an anencephalic fetus after autopsy. *Source: Charon P. 2002*.

Original author's abbreviations stand for, mandible (A), teeth (B), maxilla (C), palatines (D), "hérisséal" (E), vomer (G), nasal (H), horns (I,J), frontal (K), lacrimal (L), zygomatic (M), squamous temporal (N), petrous (P), tympanic ring (Q), squamous temporal (R), interparietal (S), parietal (T), supra-occipital (U), pars lateralis (V), basion (X), sphenoid body (Y), lesser wings of the sphenoid (Z), incus (Æ), "ingrassial (Œ), "interparietal ?" (Θ)

(2001) paraphrased the son of Étienne Geoffroy Saint-Hillaire, Isidore Geoffroy Saint-Hillaire's response (1847) to his father's inspection of the mummy:

> "He found the anencephaly case among baboon mummies, recognizing as human immediately because of his work on the subject. The infant had been mummified in the posture-hips and knees flexed, hands on knees-in which baboons are mummified, and it was furnished with a terra cotta amulet of the baboon deity. Even though it had been born to a woman, it was understood to be animal, and hence excluded from ordinary human burial. However, since it was a sacred animal, religion required that its remains be preserved in a pious fashion. The embalmer had followed the normal mummification practice and attempted to extract the brain by means of an opening through the nose, failing to recognize the significance of deformities in the posterior region of the head and neck" (18).

Saint Hillaire published a lithograph (1826) of the Hermopolis mummy from the front, rear, and right side (Figure 3.2). This picture has been crudely redrawn (Figure 3.3) several times

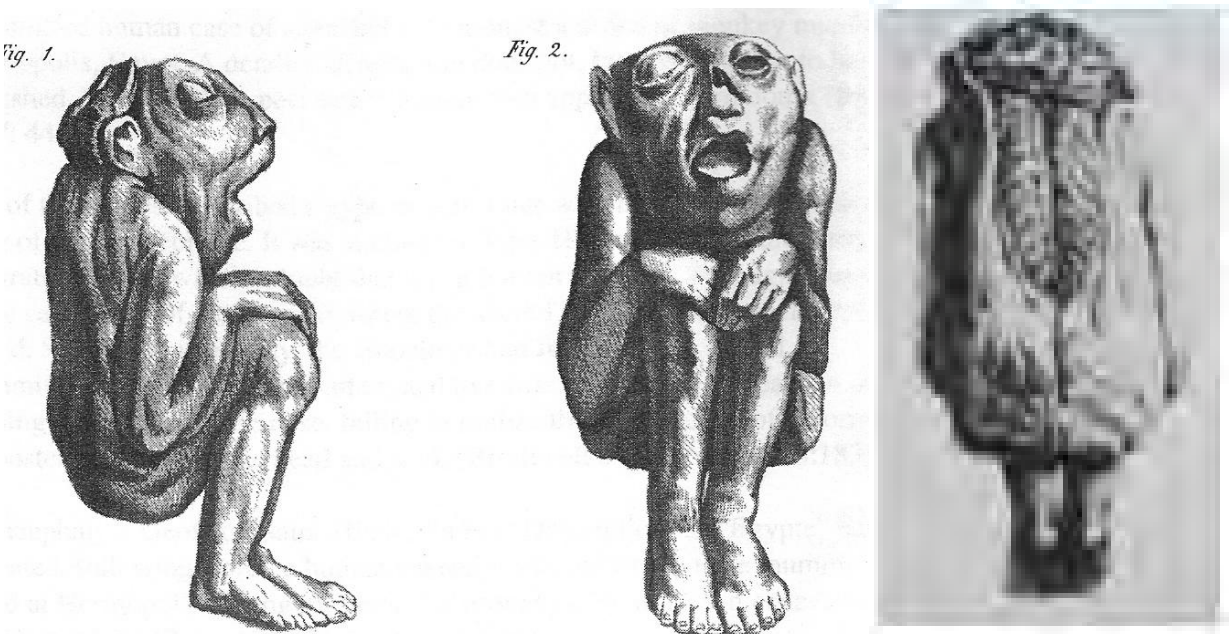


Figure 3.2: Original lithograph of the Hermopolis mummy. Most texts that refer to this individual only include the side and frontal view (left and center). To show that the Hermopolis mummy also had craniorachischisis, not just anencephaly, it is imperative that a better digital image be scanned of the rear view (right). *Source: Collins Cook D. 2002; Charon P. 2005.*

without much explanation (Brothwell and Powers, 1968; Barnes, 1994; Miller and Simon 2001) because the original publication has yet to be located. From the original depiction of the Hermopolis mummy, it appears that the defect extended from the cranial vault through the foramen magnum to the thoracic vertebrae, which is consistent with craniorachischisis, not just anencephaly. The whereabouts of the Hermopolis mummy are still unknown, although it is speculated to be amongst the collection of animal mummies in the Museum of Avignon (Collins Cook, 2002).

Figure 3.3: A crude redrawing of Etienne Geoffrey Saint-Hillarie's lithograph of the Hermopolis mummy as redrawn in Brothwell and Powers, 1968; Barnes, 1994; Miller and Simon 2001

Materials and Methods

The focus of this study, K2 B563 (Figure 3.4), was excavated from the Kellis 2 cemetery (Figure 3.5), located east of the ancient village of Kellis in the Dakhleh Oasis, Egypt (Figure 3.6). As of December 2007, a total of 700 individuals have been excavated (Dupras, pers. comm.), of which 643 adults and 78 fetal/infant burials have been analyzed (Tocheri et al., 2005). Analysis of this individual includes estimation of age and pathology. Initial pathological analyses indicated that B563 had severe cranial developmental deformations reminiscent of a neural tube defect. It is important to note that during the course of analysis the missing portion of the cranial bones were determined to be due to developmental errors and not damage due to the fact that none of the bones display any characteristics of postmortem fracture. Because there is little to no published qualitative data regarding the skeletal manifestation of neural tube defects, it was then therefore necessary to make qualitative comparisons to skeletally normal individuals and to a documented skeletal collection that contained individuals previously diagnosed with such defects.

Figure 3.4: Burial 563 during excavation.

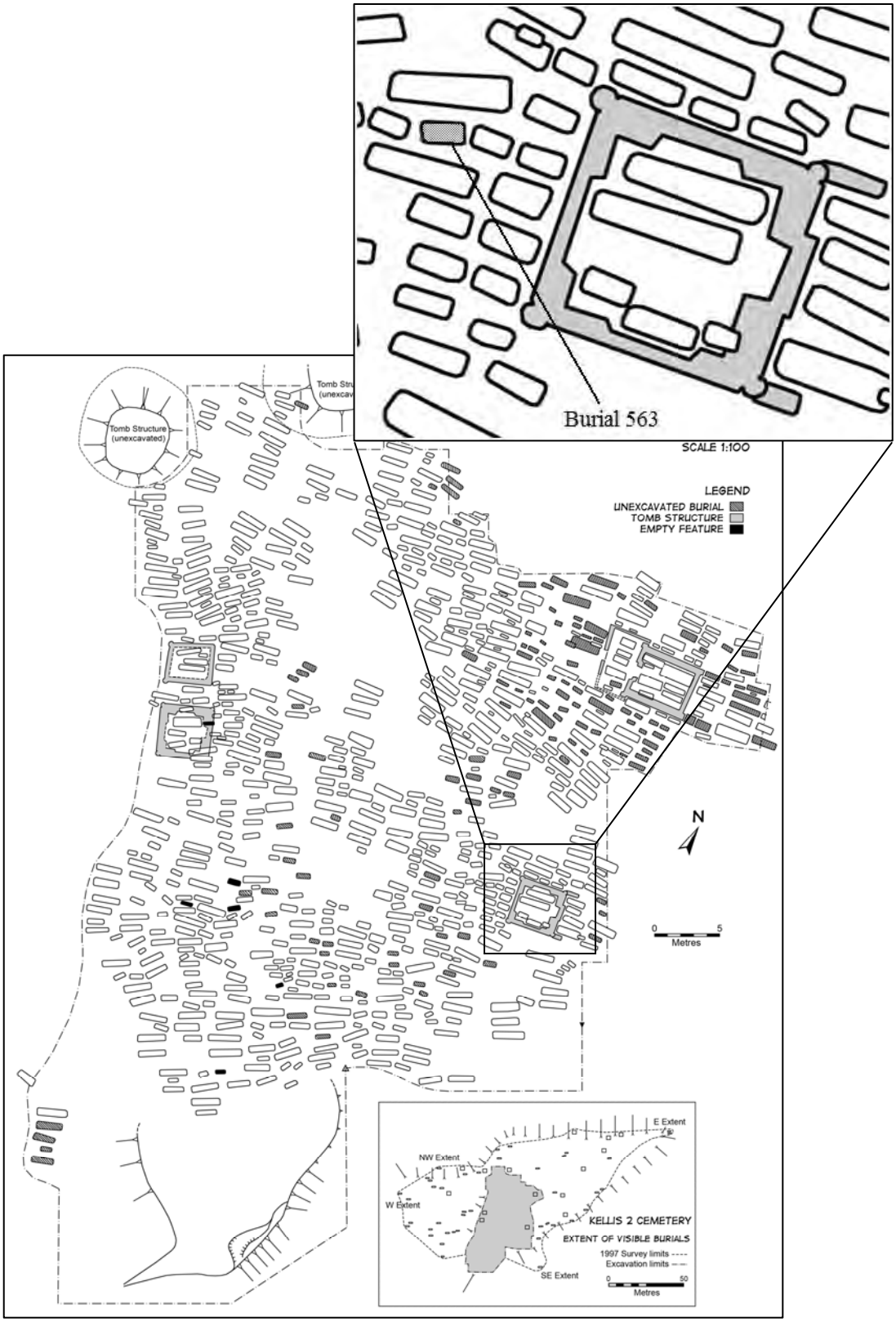


Figure 3.5: Kellis 2 cemetery burial excavations through December 2007. The inset is a close up of tomb 3 and its surroundings. Burial 563 is highlighted.

Map courtesy of S. Wheeler, University of Western Ontario.

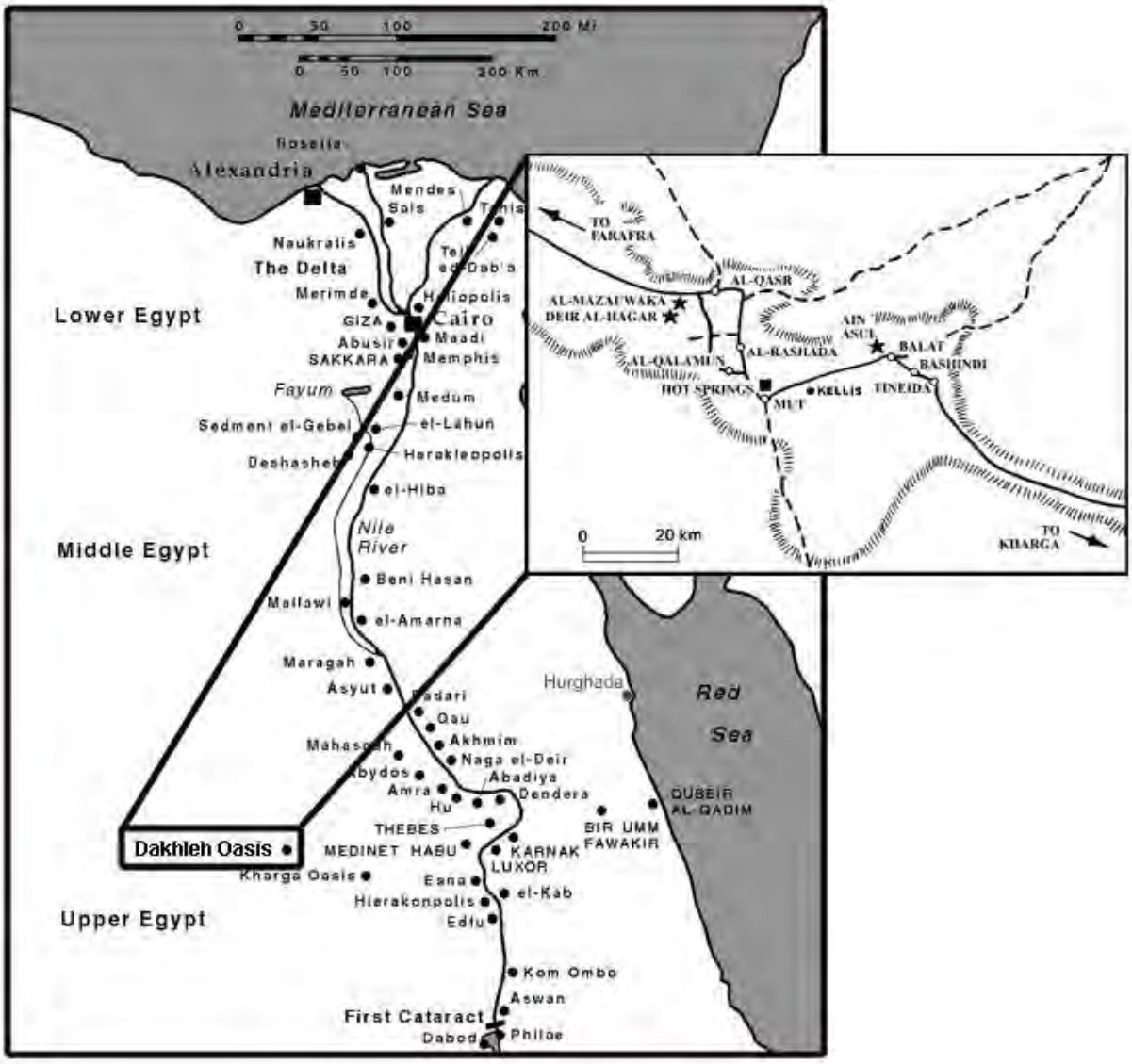


Figure 3.6: Map of Egypt. The inset denotes the Dakhleh Oasis and the location of the site of Kellis.

The collection used for comparison is housed at the Smithsonian Institution's National Museum of Natural History (NMNH) in Washington, DC. The material reviewed included both non-pathological fetal remains as well as pathologic specimens (Table 3.1). These remains were

Table 3.1: Individuals included in NMNH sample plus the individual from the Kellis 2 cemetery. Information included is all that is provided to researchers about the individuals in the NMNH collection.

CAT#	Collector	Sex	Age	Ethnicity	Notes Provided
218141	Lamb, DS	Male	06	White	About 6.5 Months, Anencephaly, Craniorachischisis, Spina Bifida
218142	Lamb, DS	Female	07	White	About 7 Months, Pseudencephalus, Spina Bifida
218143	Lamb, DS	Male	09	White	At term, Anencephaly
218144	Lamb, DS	Female	06	Mixed	About 6 months
218146	Lamb, DS	Unknown		White	Anencephaly
219199	Lamb, DS	Female		White	About 7 Months, Anencephaly, Spina Bifida, Craniorachischisis
219200	Lamb, DS	Male	09	White	Apparently at term, Anencephaly
220182	Lamb, DS	Female		White	
222089	Lamb, DS	Female		White	Twins with 222090
224710	Scurlock, HC	Male		Black	Brain Saved (224709)
224741	Morgan, EL	Male		White	
224855	Mall, FP	Male		Mixed	Premature, About 8 months, Died 3 days after birth
224859	Mall, FP	Female		Mixed	Full term, New Born, Died 6 days after birth
224884	Mall, FP	Male		Mixed	Anencephalic
224885	Mall, FP	Female		Mixed	
224888	Morgan, EL	Female	07	Mixed	About 7-8 months
226550	Hrdlička, A	Female		White	
226553	Hrdlička, A	Unknown		Unknown	Anencephalic
228369	Poole, BG	Male	06	White	About 6-7 months, Brain Saved (228368)
228839	Mall, FP	Female		White	
249557	Mall, FP	Male	09	White	New Born
250421	Buchanan, WR	Male	04	White	4.5 Months, Brain Saved (250422)
253843	Lane, TF	Male	04	White	4 Months Premature Birth
253850	Lane, TF	Female		White	
253851	Lane, TF	Male		White	
253858	Lane, TF	Female	03	Black	3 Months
271013	n/a	Unknown		Unknown	
290932	Smith, JH	Male		White	Anencephalic
299234	Smith, JH	Female		White	
299308	Holmes, JH	Male		White	
299312	Holmes, JH	Male		White	
299443	Holmes, JH	Female		White	
299453	Holmes, JH	Male		Black	
381034	Holmes, JH	Unknown	04	Unknown	4.5 Months
T007	n/a	Unknown		Unknown	
K2 B563	UCF	Unknown		Unknown	Excavated from the Dakhleh Oasis, Egypt. Pathological.

collected during the late 19th and early 20th century from lower socioeconomic groups in the Balitmore/DC area, and curated by Ales Hrdlička between 1903 and 1917 (Gindhart, 1989). Today the collection consists of nearly 400 individuals, aged from 7 weeks gestation through 10 months after birth. The majority of the collection resulted from spontaneous abortion; therefore a bias is represented in anomalous and/or pathologic remains (Huxley, 2005). Unfortunately, scant medical records exist for both the individual and its mother, and listed age estimations at the time were based on cephalic circumference, in conjunction with torso and appendage length. Hrdlička noted the information available (Gindhart, 1989), which included sex, estimated age, and some randomized external measurements. At the time of data collection the NMNH had 9 anencephalic skeletons available for examination.

Altogether 35 fetuses from the NMNH, 9 anencephalic and 26 non-pathological, were compared to the skeleton from the Dakhleh Oasis, Egypt. Of the NMNH sample, 16 were male, 14 were female, and the sex of 5 are unknown (Figure 3.7). Twenty-two of these were listed as white, 3 as black, 6 as colored, mixed or mulatto, and the ethnicity of 4 are unknown (Figure 3.8).

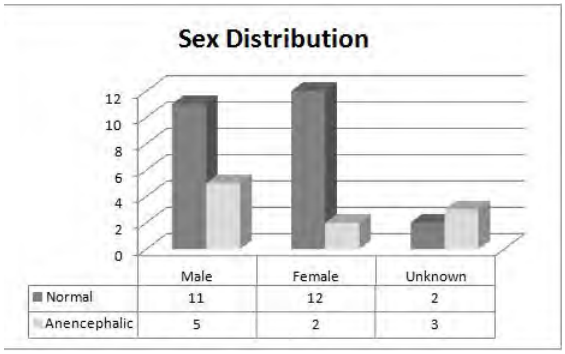


Figure 3.7: The total sex distribution of the sample including all individuals, pathologic and non-pathologic, from the NMNH plus K2 B563.

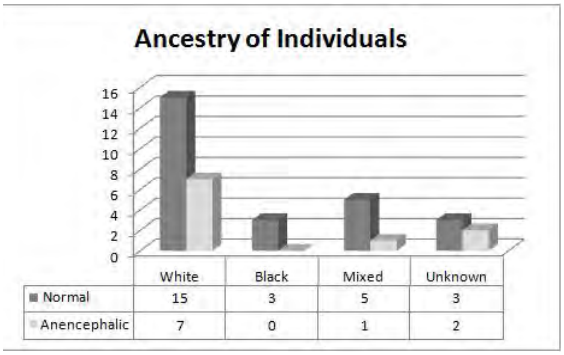


Figure 3.8: Ancestry of the total sample including all individuals, pathologic and non-pathologic, from the NMNH plus K2 B563.

Digital sliding calipers were used to take 12 cranial and post-cranial measurements (Table 3.2). Measurements were recorded to the nearest hundredth of a millimeter in order to assess age. These measurements were then compared to the data published by Scheuer and Black (2000, 2004) to provide an age estimate. Sherwood and colleagues (2000) demonstrated that that fetuses with anencephaly were over aged by 1 to 2 weeks, which was offset by the average difference of two weeks between gestational age (or weeks since the mother's last normal menstrual period) and time since conception. Based on this research it is possible that the estimation of age in the anencephalic individuals could be underestimated by one to two weeks.

Table 3.2: Measurements taken from all individuals included in NMNH sample and K2 B563. All measurements taken were from digital calipers and are based on descriptions provided by Scheuer and Black (2000).

Bone	Measurement	Description
Basion	Length	Midsagittal distance from anterior border of foramen magnum to anterior border
Basion	Width	Maximum width at level of lateral angles
Zygomatics	Width	Medial end of infra-orbital border to superior end of frontal process
Clavicle	Length	Maximum length of the entire bone
Scapula	Height	Distance between the superior and inferior angles
Scapula	Width	Distance between margin of glenoid fossa and medial end of spine
Humerus	Length	Maximum length of the entire bone
Ulna	Length	Maximum length of the entire bone
Radius	Length	Maximum length of the entire bone
Femur	Length	Maximum length of the entire bone
Tibia	Length	Maximum length of the entire bone
Fibula	Length	Maximum length of the entire bone

Results

Qualitative analysis of the anencephalic individuals from the NMNH collection revealed specific anatomical abnormalities associated with anencephaly. The anomalies of similarly aged fetuses (Figure 3.9) are consistent in comparison with those displayed by K2 B563 (Table 3.3).

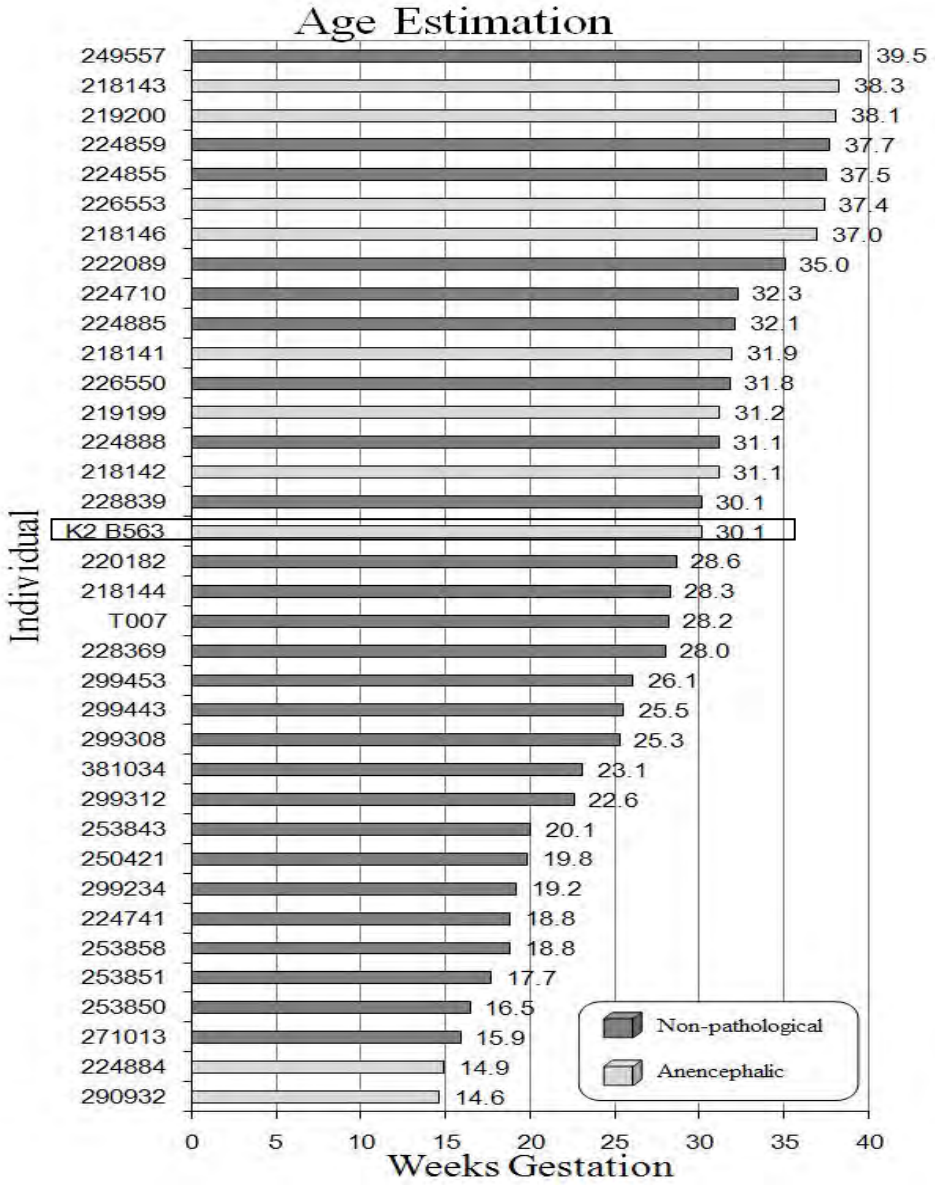


Figure 3.9: Estimated age in weeks gestation for the total sample from the NMNH plus K2 B563. The focus of this study, K2 B563 has been circled.

Table 3.3: Comparison of cranial bones showing the difference in morphology between normal development and anencephaly. Note that K2 B563 displays anencephalic traits in each bone.

Cranial Bones	Normal	Anencephalic	K2 B563
PARIETALS	2 unfused elements. Eliptical in shape and convex exteriorly with serrated borders.	Complete absence of bone.	Complete absence of bone.
OCCIPITAL	4 unfused elements.	Variable number of elements.	4 unfused elements.
Squamous	Shallow bowl shaped appaearance with a prominent external occipital protuberance at the junction	If any bone from squamous is present it is unfused and uncharacteristic in shape.	No unified squama.
pars interparietalis	Fan shaped with thin feathery borders.	Complete absence of bone.	Complete absence of bone.
pars supra-occipitalis	Wedge shaped with thick serrated borders.	If this bone does ossify it is undersized. Robust. Appears as a small wedge or sickle shape with a feathered process extending medially and superiorly.	One half is sickled shaped and very robust. The whereabouts of the other half are unknown.
Pars Lateralis	A thickened medial border and a wedge shaped posterior border. 2 inferior oimbs extends medially to create the hypoglossal canal.	Fairly similar in shape to normal fetuses, although the bones are more robust and narrow.	Fairly similar in shape to normal fetuses, although the bones are more robust and narrow.
Pars Basilaris	Robust and square with a D-shaped out out at the superior surface. The inferior surface is slightly concave. Wider than it is long.	Similar in shaper to normal fetuses although the proportions are inverted: the bone is longer than it is wide.	Similar in shaper to normal fetuses although the proportions are inverted: the bone is longer than it is wide.
TEMPORALS	2 unfused elements on either side	Early fusion of all elements.	Early fusion of all elements.
Petrous	Cilindrycal in shape. Medially the internal acoustic meatus is anterior to the large subarcuate fossa. Superiorly the surface is smooth.	Highly irregular in shape and shorter in total length than normal. Foramen appear smaller and are located irregularly.	Highly irregular in shape and shorter in total length than normal. Foramen appear smaller and are located irregularly.
Squamous	A delicate, nearly flat, semicircular plate with serrated borders. The zygomatic process extends anteriorly.	Appears to have curled back upon itself onto the petrous portion. The zygomatic process is the only portion that is recognizable, yet is shortened.	Appears to have curled back upon itself onto the petrous portion. The zygomatic process is the only portion that is recognizable, yet is shortened.
Tympanic Ring	A nearly complete circle of bone in the shape of a ring fused to the squamous.	Fuses early. Appears more elliptical than circular.	Fuses early. Appears more elliptical than circular.
FRONTALS	2 unfused elements. An irregular, yet bowl shaped bone. The superior squamous portion is rounded and convex exteriorly. The orbital plate meets the squamous inferiorly with a dome shape.	Complete absence of the squamous portion. Only the anterior border of the orbit forms, with an excessive curvature. The shape of this bone is the most varible in anencephalics.	Complete absence of the squamous portion. Only the anterior border of the orbit forms, with an excessive curvature. The superior border forms a right angle medially.
SPHENOID	3 unfused elements	Early fusion of all elements.	Early fusion of all elements
Body	Cube shaped with 2 alar processes which resemble feathers.	Far more narrow than normal. Almost rounded.	Far more narrow than normal. Almost rounded.
Lesser wings	Flat and arrow-head shaped bone which fuses to the body.	Nearly unrecognizable. Curled back upon the body. Severely undersized.	Nearly unrecognizable. Curled back upon the body. Severely undersized.
Greater wings	Triangular in shape this bone's lateral $^2/_3$ are thin and slightly concave anteriorly. The medial $^1/_3$ is a complex root for attaching to the body.	Fused to the body early. The malformations are nearly indescribable. The root is wider and more robust whereas the wing is also curcle back towards the body.	Fused to the body early. The malformations are nearly indescribable. The root is wider and more robust whereas the wing is also curcle back towards the body.
ZYGOMATICS	A robust bone resembling an upside down letter T. The superior process extends towards the frontals, the anterior process extends towards the maxilla, and the posterior process extends toward the temporal.	Appears normal with the exception of the complete or nealry complete absence of the temporal process.	Appears normal with the exception of smaller temporal process. More robust than normal.

During analysis it was noted that most of the 9 anencephalic fetuses from the NMNH were missing both parietals; K2 B563 is also missing both parietals (Figure 3.10).

The NMNH anencephalics were also missing the squamous portion of the occipital, or only a minor fragment of the supra-occipitalis was present. The squamous portion of K2 B563's occipital was missing, although a thickened wedge shaped portion of the supra-occipitalis was

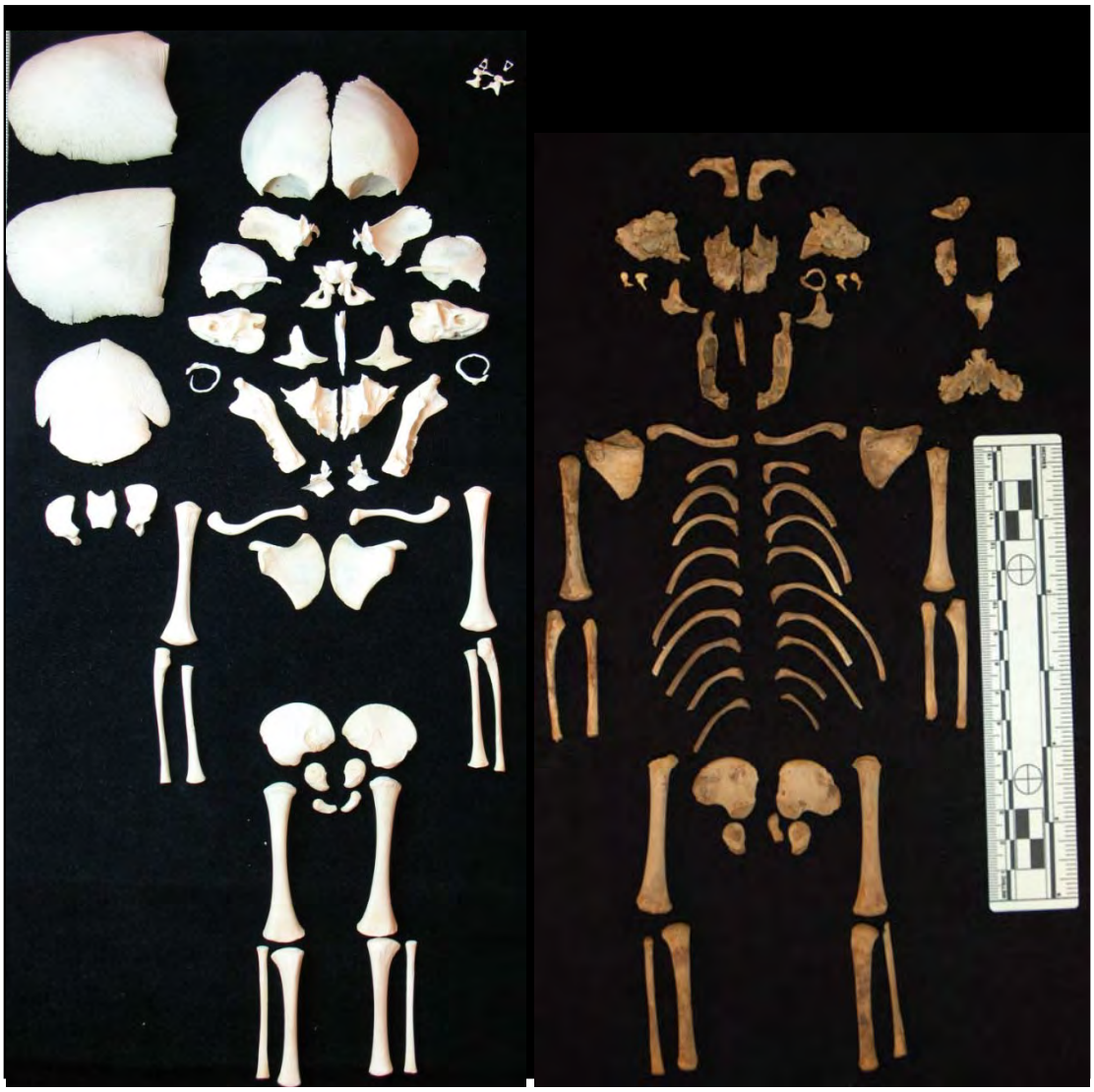


Figure 3.10: A nonpathological skeleton (left) compared to K2 B563 (right). Note: several bones were excluded from the photo including the vertebrae, ribs (left), as well as the bones of the hands and feet. Note the state of preservation of K2 B563 and the completeness of the postcranial skeleton.

present. Furthermore, the NMNH collection of anencephalic fetuses' pars lateralis and pars basilaris were malformed. The shape of this anomaly varied between the 9 individuals analyzed. These bones of the occipital in K2 B563 were also abnormal in that they were longer and narrower than in normal fetuses (Figure 3.11).

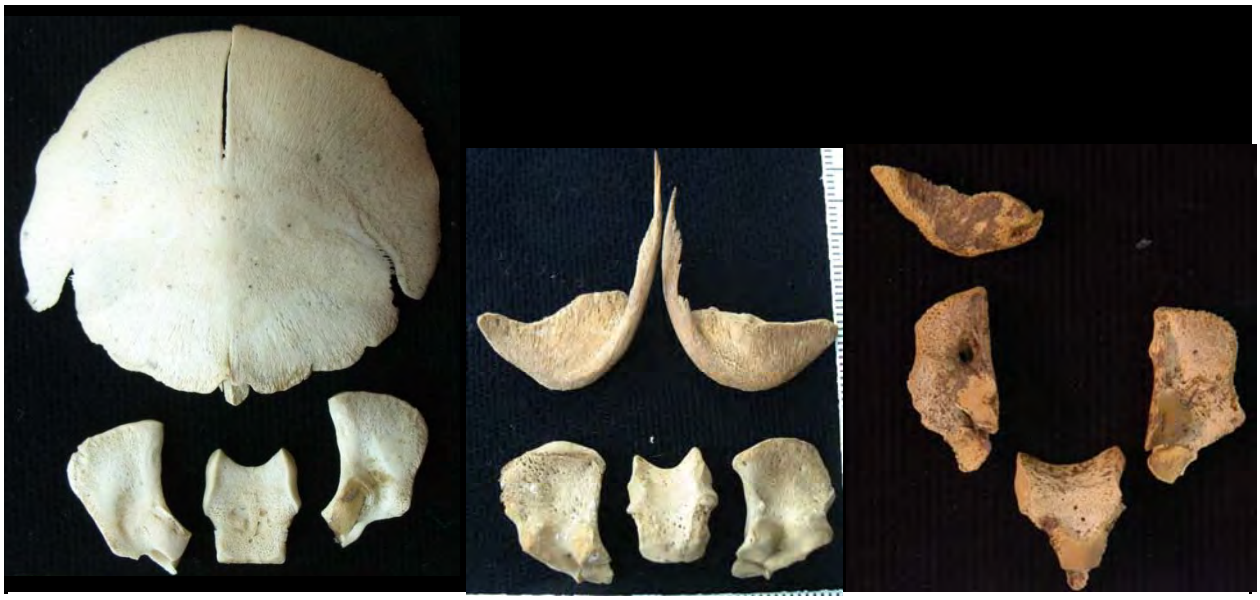


Figure 3.11: Comparing the occipital bones of a nonpathological fetus (left), a clinical anencephalic (center), and K2 B563 (right).

In the NMNH collection the anencephalic frontal bones only exhibit the portion that makes up the superior portion of the eye orbit, and are completely missing the squamous portion. When compared to one of the NMNH anencephalic skeletons of the same age, the malformed frontals of K2 B563 were identical in morphology (Figure 3.12).

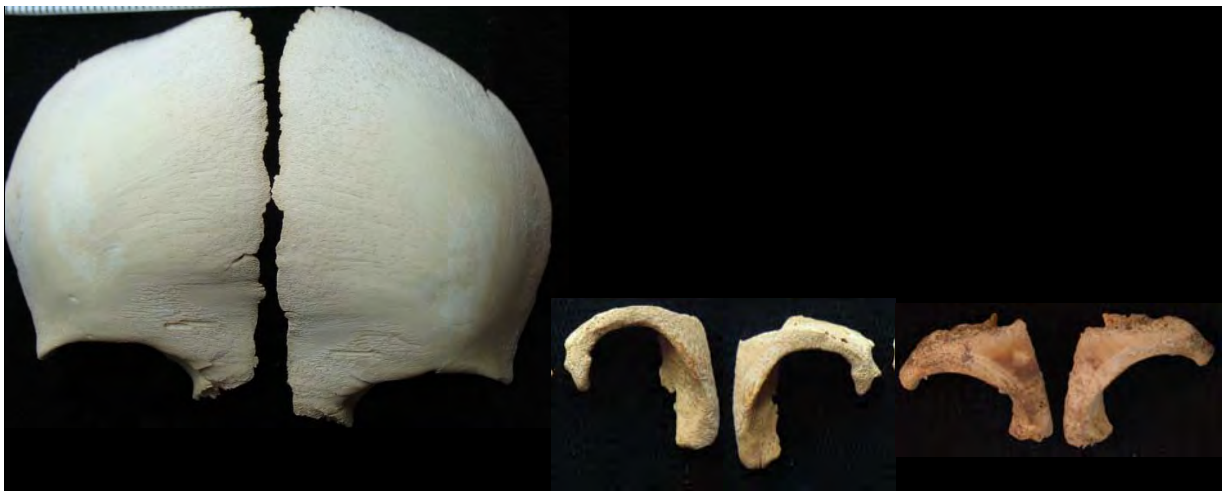


Figure 3.12: Comparing the frontal bones of a nonpathological fetus (left), a clinical anencephalic (center), and K2 B563 (right).

In normal development, the temporals are comprised of three elements which fuse to form a single bone shortly after birth. The NMNH remains show that the temporals of these anencephalic individuals fused prematurely, sometime after 14 weeks and before 29 weeks gestation. Even though the three elements fuse early, the squamous portion of the temporal is nearly missing; the only feature that remains is a shortened zygomatic process. These findings for the temporals are consistent with the morphology of the temporals of K2 B563 (Figure 3.13).

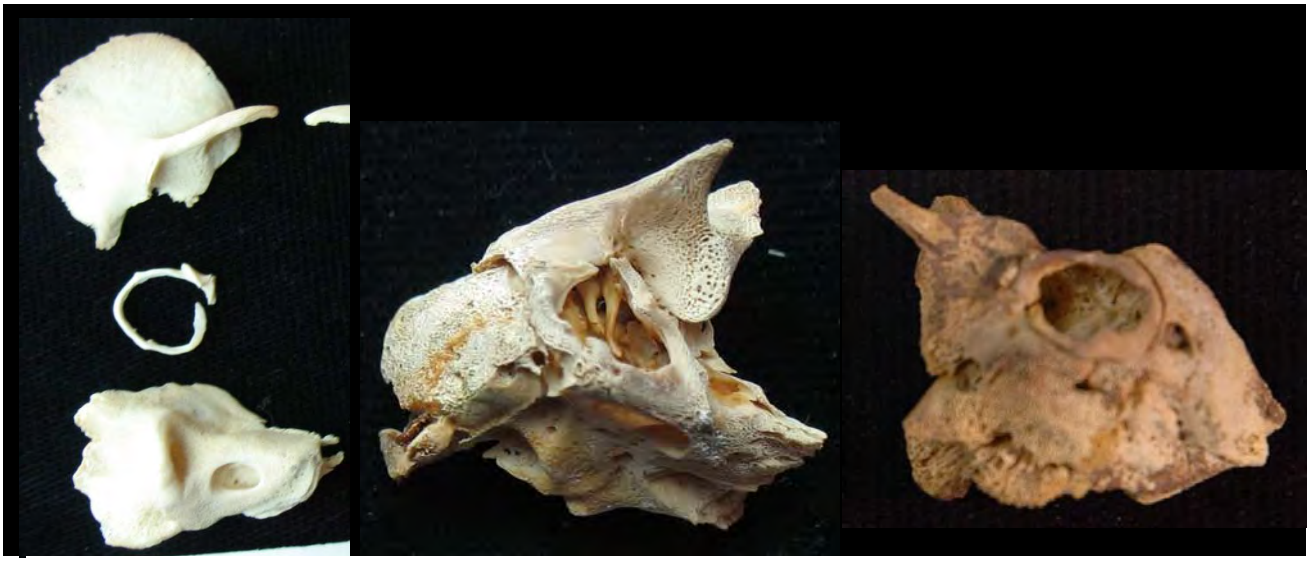


Figure 3.13: Comparing the right side temporal bone of a nonpathological fetus (left), a clinical anencephalic (center), and K2 B563 –left side (right).

The temporal processes of the NMNH anencephalic zygomatics were either missing altogether, or were shortened and broad. The temporal process of K2 B563's zygomatics were similar the NMNH collection in that the temporal process was far shorter and much broader than it ought to be for its estimated age (Figure 3.14).

Figure 3.14: Comparing the zygomatics of a nonpathological fetus (left), a clinical anencephalic (center), and K2 B563 (right).

The most deformed of the anencephalic bones in the NMNH collection is the sphenoid. This bone is comprised of a body, two lesser wings, and two greater wings. During normal development, the lesser wings fuse to the body during the 20th week of gestation, and the greater wings fuse to the body during the first year of life. All of the NMNH anencephalic fetuses' lesser wings fused prematurely and were so malformed that they were nearly unrecognizable. Most of these fetal skeletons' greater wings had also fused prematurely, and all of the greater wings are severely misshapen. When compared to one of the NMNH anencephalic skeletons of the same age, the malformed sphenoid of K2 B563 was identical in morphology (Figure 3.15).

Figure 3.15: Comparing the sphenoid of a nonpathological fetus (left), a clinical anencephalic (center), and K2 B563 (right).

During the analysis of the measurements taken on the craniofacial elements of the anencephalics at the NMNH it became clear that this NTD highly distorted the approximated age for each individual. The age estimate, in weeks gestation, based on the measurement of each bone can be seen in Figure 3.16. Here you can see, for example, that anencephalic individual 218142's length of the basion suggests 38 weeks gestation, whereas the width of the same bone suggests an age of 25 weeks gestation. This trend of drastic variation in age estimation of the anencephalic's craniofacial skeletal measurements varies amongst the 10 individuals. Not all individuals from this sample have such drastic variation in age. It is being suggested that this is caused by a range in severity in anencephaly. Therefore, the more severe the defect the more drastically the measurements used to estimate age are varied.

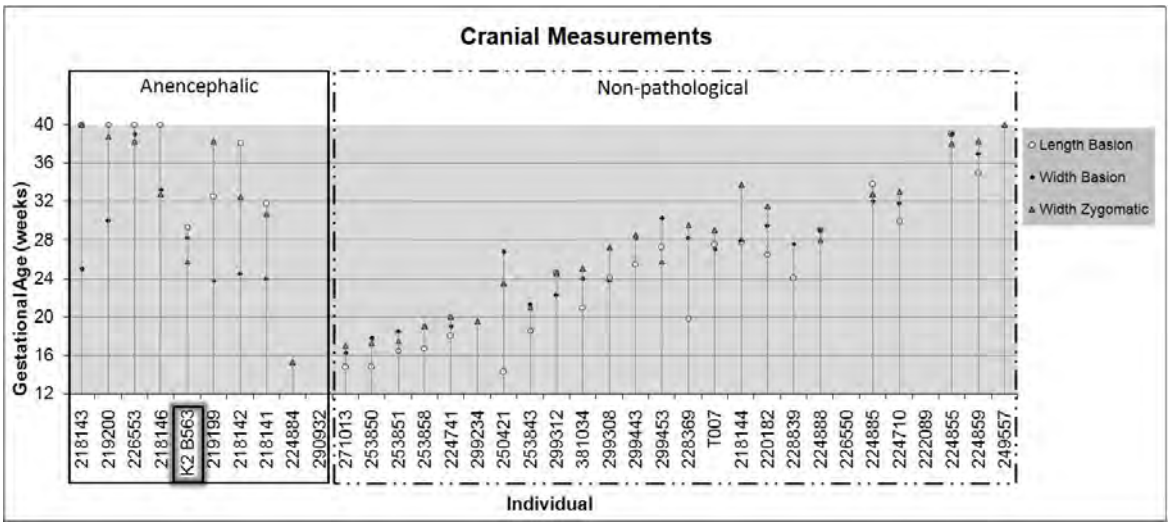


Figure 3.16: Comparing the age estimations based on cranial measurements of anencephalic fetuses (left), to non-pathological fetuses (right). The focus of this study, K2 B563, is circled.

When directly comparing the age estimates based on cranial measurements in the non-pathological specimens very few displayed this drastic range of variation. This variation in these few individuals may indicate a currently unknown pathology which causes the bones to be malformed, or it could simply be an error in measurement. Overall, when the cranial age estimations are analyzed alongside the appendicular age estimations, which can be seen in Figure 3.17, little variation in age is seen in both the nonpathological individuals and the anencephalics with the exception of those with craniorachischisis (226553A, 219199, and 218141). This evidence further supports the normal development of the postcranial skeleton in individuals with the NTD anencephaly.

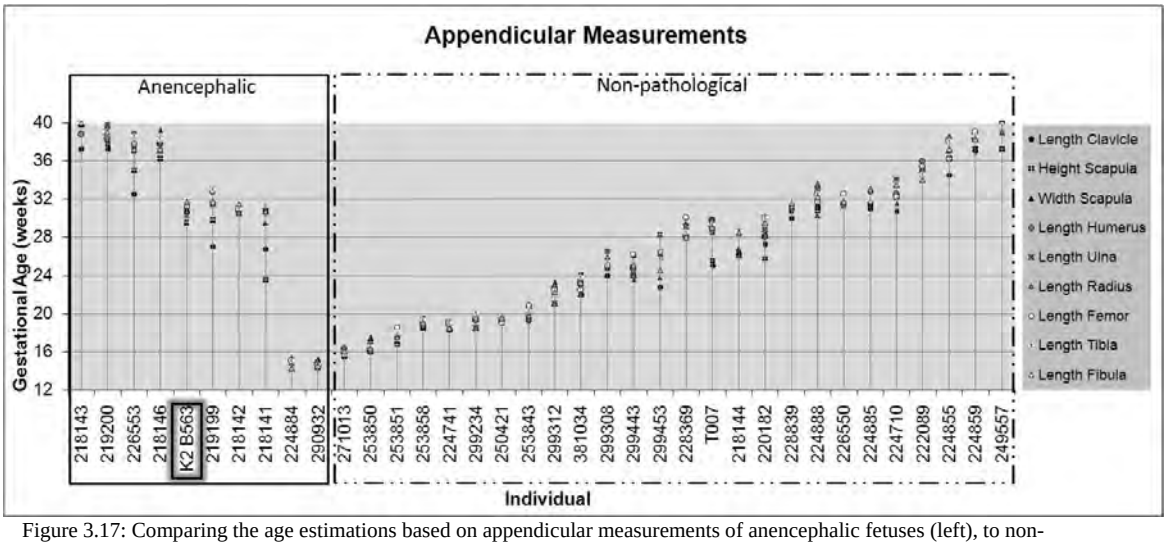


Figure 3.17: Comparing the age estimations based on appendicular measurements of anencephalic fetuses (left), to non-pathological fetuses (right). The focus of this study, K2 B563, is circled.

Discussion and Conclusion

Based on the morphological comparison of the documented anencephalics from the Smithsonian Institution National Museum of Natural History to that of the individual from Kellis 2 in the Dakhleh Oasis, Egypt, K2 B563 the osseous representation of the cranium can be attributed to the neural tube defect anencephaly. It is worth reemphasizing that the arid climate of the Egyptian desert is extremely conducive to the superb preservation of skeletal material. This phenomenon is seen not only in this fetus, but throughout the Kellis 2 cemetery. Also there is no evidence of fracture, whether peri- or postmortem, present in K2 B563 which rules out the possibility that the cranial bones were damaged during the process of excavation.

When comparing each cranial element of K2 B563 to the observations from the NMNH collection, the similarities in morphology to the anencephalics were consistent. Although it could have been possible that the parietals of K2 B563 were never recovered, the evidence based on a lack of parietals in the nine anencephalic individuals from the NMNH collection support the theory that they had never formed, not that they had been overlooked during excavation. The missing interparietalis portion of the occipital could have initially been attributed to poor excavation technique, but as in the parietals, this bone is not at all represented in the nine anencephalic individuals from the NMNH collection. The presence of the unusually shaped supra-occipitalis, pars lateralis and pars basilaris also correspond to the physical representation of the occipital bone of the comparative individuals from the NMNH collection. Having a pair of nearly identical frontals from a documented anencephalic fetus further supports the manifestation of this NTD in K2 B563. The early fusion of the temporals and the lack of its squamous portion,

with the exception of the shortened zygomatic process, seen in K2 B563 was also noted in almost all of the NMNH's anencephalic individuals. The articulating process on the zygomatic that stretches to the temporal in K2 B563, as well as in the anencephalic individuals at the NMNH, was far shorter than those without pathologic conditions. Again, there was no physical evidence among the NMNH anencephalic collection or in K2 B563 that either process on the temporal or the zygomatic had been broken therefore causing the shortened length. As with the frontals, the near identical comparison in morphology of the severely malformed sphenoid from a documented anencephalic fetus further supports the manifestation of anencephaly in K2 B563.

When the analysis of K2 B563 is compared to the description of the Hermopolis mummy, the only published archaeological anencephalic, numerous morphological similarities are evident; the only differences arise due to the Hermopolis mummy having craniorachischisis in addition to anencephaly. Now having two examples of anencephaly in the archaeological record, both from Egypt, can now allow bioarchaeologists to hypothesize about the evolution of cultural perceptions of deformed children. Hopefully, more anencephalic individuals will be discovered in Egypt that date between the Old Kingdom and Roman rule, so that bioarchaeologists can gain a better understanding of the perception and funerary practices of babies born with severe birth defects.

It is fair to say that it is conclusive that the fetal skeleton K2 B653 from the Dakhleh Oasis, Egypt, does in fact display the classic skeletal indicators of the neural tube defect anencephaly. When compared to the 9 other clinical skeletons in the NMNH collection, the evidence provides support that this archaeological specimen has formed these bony

characteristics due to the neural tube defect anencephaly. Nothing supports that these bones have been broken, or have disintegrated over time due to natural processes.

References

Baker BJ, Dupras TL, and Tocheri MW. 2005. *The Osteology of Infants and Children*. College Station: Texas A&M University Press.

Barnes E. 1994. *Developmental Defects of the Axial Skeleton in Paleopathology*. Niwot: University Press of Colorado.

Birrell M. 1999. Excavations in the cemeteries of Ismant el-Kharab. In: *Dakhleh Oasis Project: preliminary reports on the 1992-1993 and 1993-1994 field seasons,* Hope CA, Mills AJ (eds.) Oxford: Oxbow. 43-59.

Bowen GE. 2003. Some observations on the Christian burial practices at Kellis. In: *The Oasis Papers III: The Proceedings of the Third International Conference of the Dakhleh Oasis Project,* Bowen GE, Hope CA (eds.) Oxbow: Oxford; 166-182.

Brothwell DR, and Powers R. 1968. Congenital Malformations of the Skeleton in Earlier Man. In: *Symposia of the Society for the Study of Human Biology,* Volume VIII. New York: Pergamon Press. p 173-203.

Charon P. 2002. Paleopathology Queries and Comments. *Paleopathology Newsletter* 119:11-19.

Collins Cook D. 2001. Neglected Ancestors: Etienne and Isidore Geoffroy Saint-Hillaire. *Paleopathology Newsletter* 116:17-20.

Collins Cook D. 2002. …and the Mystery Solved? *Paleopathology Newsletter* 119:19.

Dambska M and Wisniewski KE. 1999. *Normal and Pathologic Development of the Human Brain and Spinal Cord*. London: John Libbey & Company Ltd.

Dolk H, De Wals P, Gillerot Y, Lechat MF, Ayme S, Cornel M and Cuschieri A. 1991. Heterogeneity of Neural Tube Defects in Europe: The Significance of Site Defect and Presence of Other Major Anomalies in Relation to Geographic Differences in Prevalence. *Teratology*. 44:547-559.

Dupras TL and Schwartz TP. 2001. Strangers in a Strange Land: Stable Isotope Evidence for Human Migration in the Dakhleh Oasis, Egypt. *Journal of Archaeological Science* 28: 1199-1208.

Dupras TL and Tocheri M. 2007. Reconstructing infant weaning histories at Roman period Kellis, Egypt using stable isotope analysis of dentition. *American Journal of Physical Anthropology* 134:63-74.

Fairgrieve SI, and Molto JE. 2000. Cribra Orbitalia in Two Temporally Disjunct Population Sample from the Dakhleh Oasis, Egypt. *American Journal of Physical Anthropology* 111(3):319-332.

Geoffroy Saint-Hillaire E. 1826. Note about an Egyptian monster found in the ruins of Thebes, in Egypt, by M. Passalascqua. *Bulletin des Sciences Medicales* VII: 105-108.

Geoffroy Saint-Hillaire I. 1847. Vie, Travaux et Doctrine Scientifique d' Étienne Geoffroy Saint-Hillaire par son Fils. Paris: Bertrand.

Green NDE and Copp AJ. 2006. The Embryonic Basis of Neural Tube Defects. In: Diego Wyszynski, editor. *Neural Tube Defects*. New York: Oxford University Press p15-28.

Gindhart PS. 1989. An early twentieth-century skeleton collection. *Journal of Forensic Sciences*. 34(4):887-893.

Hope CA. 2001. Observations on the dating of occupation at Ismant el-Kharab. In *The Oasis Papers I: The Proceedings of the First Conference of the Dakhleh Oasis Project,* Marlow CA, Mills AJ (eds.) Oxbow: Oxford; 43-59.

Huxley AK. 2005. Gestational Age Discrepancies Due to Acquisition Artifact in the Forensic Fetal Osteology Collection at the National Museum of Natural History, Smithsonian Institution, USA. *American Journal of Forensic Medicine & Pathology*. 26(3):216-220.

Marlow CA. 2001. Miscarriages and Infant Burials in the Dakhleh Cemeteries: An Archaeological Examination of Status. In *The Oasis Papers I: The Proceedings of the First Conference of the Dakhleh Oasis Project,* Marlow CA, Mills AJ (eds.) Oxbow: Oxford; 105-110.

Miller E, and Simon K. 2001. Anencephaly – Something Missing from the Archaeological Record? *Paleopathology Newsletter* 115:9-11.

Mills AJ. 1984. Research in the Dakhleh Oasis. In *Origin and Early Developemt of Food Producing Cultures in North-Eastern Africa,* Krzyzaniak L, Kobusiewicz M (eds). Polish Academy of Sciences: Poznan; 205-210.

Molto JE. 2001. The comparative skeletal biology and paleoepidemiology of the people from 'ein Tirghi and Kellis, Dakhleh Oasis, Egypt. In *The Oasis Papers I: The Proceedings of the*

First Conference of the Dakhleh Oasis Project, Marlow CA, Mills AJ (eds.) Oxbow: Oxford; 81-100.

Molto JE. 2000. Humerus Varus Deformity in Roman Period Burials from Kellis 2, Dakhleh, Egypt. *American Journal of Physical Anthropology* 113(1):103-111.

Moore CA. 2006. Classification of Neural Tube Defects. In, *Neural Tube Defects*. Wyszynski DF; editor. New York: Oxford University Press.

Parsche F and Zimmerman P. 1991. Results of computer assisted studies of population structure and burial practices of adults of the late Roman burial ground in the Minshat Abu Omer (east Nile delta). *Anthropologischer Anzeiger (Stuttgart)* 49:65-83.

Scheuer L, and Black S. 2004. *The Juvenile Skeleton*. London: Academic Press.

Scheuer L, and Black S. 2000. *Developmental Juvenile Osteology*. London: Academic Press.

Shaffer LG, Marazita ML, Bodurtha J, Newlin A, and Nance WE. 1990. Evidence for a major gene in familial anencephaly. *American Journal of Medical Genetics*.36:97-101.

Sherwood RJ, Meindl RS, Robinson HB, and May RL. 2000. Fetal Age: Methods of Estimation and Effects of Pathology. *American Journal of Physical Anthropology* 113:305-315.

Stewart JD, Molto JE, Reimer P. 2003. The chronology of Kellis 2: the interpretive significance of radiocarbon dating of human remains. In: *The Oasis Papers III: The Proceedings of the Third International Conference of the Dakhleh Oasis Project,* Bowen GE, Hope CA (eds.) Oxbow: Oxford; 345-364.

Tocheri M, Dupras TL, Sheldrick P, Molto JE. 2005. Roman Period Fetal Skeletons from the East Cemetery (Kellis 2) of Kellis, Egypt. *International Journal of Osteoarchaeology*. 15(5): 326.

CHAPTER 4
CONCLUSIONS

Discussion

The preservation of human fetal skeletons in the archaeological record is remarkable, and occurs fairly infrequently; the discovery of well preserved fetal remains during excavation is even more unusual. Understandably, coming across fetal skeletons that have been buried for any length of time with birth defects is an extremely rare occurrence because these skeletons are incredibly fragile and are exposed to numerous environmental threats. Often, fetal skeletons are buried in a different manner than adults, which may or may not aid in their preservation. Unfortunately, these cultural practices do not always protect fetal skeletons from total decomposition. Fetal bones are often badly damaged or even destroyed due to careless excavation technique (Barnes, 1994). For those individuals that pass the test of time, and are meticulously excavated, even then many do not find their way into modern osteological collections because they are commonly mislabeled as non-human. This unfortunate fact is particularly common when only fragmentary remains are recovered or if the bones originated in an area with poor preservation (Scheuer and Black, 2000).

This project has the potential to educated bioarchaeologists in identifying developmental defects for what they are, and not as postmortem trauma. The reason for this research was to not only rule out damage as the cause of the missing portions of K2 B563's skull, but also to be able to diagnose neural tube defects in archaeological skeletons. As a result of the comparative

analysis of the nine clinical individuals from the Smithsonian Institution National Museum of Natural History in Washington D.C. and K2 B563 from the Kellis 2 cemetery in Dakhleh Oasis, Egypt, a foundation has been provided for further research into archaeological skeletons with neural tube defects.

An introduction to neural tube defects is covered in chapter 2, with an emphasis on anencephaly. Part of comprehending what a NTD is requires a basis of understanding in how and when errors occur during early embryological development. Although NTDs occur today less frequently, the cause it is still unknown. Previous research studies (Fields et al., 1978; Garol et al., 1978; Metzner et al., 1978) have begun to differentiate between the types of anencephaly by describing the cranial structures as a whole. As this is the only type of descriptive data published on the varying levels of severity within anencephaly, it provides a useful foundation for the in depth analysis of the skeletal elements of these individuals.

An analysis of nine documented anencephalics in a collection at the Smithsonian Institute's National Museum of Natural History was conducted to provide a description of each bone's morphology due to malformation. It was found that although the cranial structures were severely misshaped from the NTD, the post-cranial elements developed normally, with the exception of those with craniorachischisis, as the defect extends through the base of the skull and continues down the spine also affecting their scapulae. It was also found that the squamous portions of all the bones, including the parietals in their entirety, either did not form at all or were so growth stunted that they were nearly nonexistent. Also, the temporals and the sphenoid should fuse into a single element after birth; these individuals displayed early fusion.

Additional cranial bones had unique malformations as well. The bones that surround the foramen magnum, the pars lateralis and basion, were longer and far narrower than expected. Other than fusing early, the petrous portion of the temporal displayed a highly irregular shape, and the sphenoid was nearly unrecognizable. The frontals only exhibited the superior portion of the eye orbit, and the zygomatics were either missing or had stunted temporal processes. It was concluded that this data was sufficient to diagnose the presence of anencephaly in an archaeological context.

Chapter 3 introduced a fetus found in the Kellis 2 cemetery, in the Dakhleh Oasis, Egypt with severe cranial deformities. Upon excavation of this individual, K2 B563, it was thought that it displayed classic signs on anencephaly. Having no other skeleton to make a comparison to from an archaeological context, a clinical comparison was necessary. A review of NTDs was provided from the previous chapter, with the inclusion of a description of the only anencephalic ever found archaeologically, the Hermopolis mummy. Unfortunately the whereabouts of this individual is unknown, therefore a direct comparison could not be made.

The K2 B563 skeleton was thus compared to the nine documented anencephalics at the NMNH. It was found that, when laid side by side, the morphological characteristics of K2 B563's malformations were nearly identical to those from the NMNH. K2 B563 was also missing the same portions, the squamous, of all the bones just as the Smithsonian's anencephalics were. This qualitative comparison allowed for the conclusion the K2 B563 had not been damaged during excavation or that the missing pieces were due to decomposition, and that in fact the malformations present were indicative of anencephaly.

Future Considerations

Archaeological human skeletal populations provide researchers with an abundance of information regarding past cultures. In some populations an invaluable basis of paleopathological data is also available. However, there are limitations that present problems for research. Burial practices of still born fetuses vary across time and by culture and the very likelihood of excavating a population of well preserved fetal remains is low. Furthermore, the discovery of fetuses with paleopathological conditions is highly unlikely due to their susceptibility to rapid decomposition and a range of cultural practices that mandate how fetuses are to be disposed of upon death. Also, anencephaly in and of itself is a rare defect in any given population. The rate of occurrence in modern times is speculative at best, therefore the chance of discovering another individual to compare to K2 B563 is very low. Nevertheless, variation amongst the skeletal manifestations of neural tube defects, anencephaly in particular, would be a recommended topic for further research.

Paleopathology has always based its comparison of past skeletal populations on modern medical research. To fully understand to origins of K2 B563's defect more research needs to be conducted to determine if the cause of NTDs is genetic, environmental, a combination of the two, or another cause not yet investigated. Also, when a baby is delivered stillborn, or dies within a short time, it is imperative that a more in depth description of the NTD should be provided upon autopsy. Even though this description would necessarily be able to provide crucial data regarding the morphology of each bone malformed by these defects, it would be an invaluable source to bioarchaeologists and paleopathologists alike. Furthermore, a study that

pictorially documents varying levels of severity in NTDs from existing skeletal collections needs to be conducted prior to establishing specific guidelines for diagnosing NTDs from skeletal remains.

This study opens doors for other researchers who have found fetal skeletal remains in at an archaeological site that appear to display a neural tube defect, yet had previously been unidentifiable. Since diagnostic tools have now been developed for the analysis and interpretation of anencephaly, future studies should include investigations beyond the biological information provided by a skeleton, and to include interpretation of the cultural practices involving mortuary customs regarding the death of a child with severe deformities.

References

Barnes E. 1994. *Developmental Defects of the Axial Skeleton in Paleopathology*. Niwot: University Press of Colorado.

Fields HW., Metzner L., Garol JD., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses I. Cranial Floor. *Teratology*. 17:57-66.

Garol JD., Fields HW., Metzner L., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses II. Calvarium. *Teratology*. 17:67-74.

Metzner L., Fields HW., Garol JD., and Kokich VG. 1978. The Craniofacial Skeleton in Anencephalic Human Fetuses III. Facial Skeleton. *Teratology*. 17:75-82.

Scheuer L, and Black S. 2000. *Developmental Juvenile Osteology*. London: Academic Press.