that of the urine plus saliva samples from the same subjects. The specimen obtained while the patient was not being supervised was assumed to be a mixture of urine and saliva. In each case the unmixed urine had a normal or nearly normal amylase content, but the saliva plus urine specimen had a very high content.

Discussion

The signs and symptoms of acute pancreatitis are often nonspecific. Unfortunately, the laboratory confirmation of pancreatitis is imprecise; the serum and urinary amylase values may be normal in documented acute pancreatitis, and they may be elevated in conditions other than acute pancreatitis.[1,6,7] The importance of an elevated urinary amylase with a normal serum amylase in the diagnosis of acute pancreatitis is questionable.[1,8] A more reliable indicator of acute pancreatitis has been renal clearance of amylase, but this, too, is nonspecific.[8,9] Pancreatic amylase is an alpha 1 to 4 glycosidase, whereas salivary amylase is an alpha 1 to 4 amylase. Isoamylases can be separated, but this is not routinely done; most laboratories measure a combination of both isoenzymes when they assay serum or urinary amylase. In normal healthy subjects, salivary amylase accounts for 66 per cent of total serum amylase, whereas the pancreas is the source of the increased serum amylase caused by pancreatitis.[10]

Our patient was apparently aware of the similarity between salivary and urinary amylase and used this knowledge to elevate his urinary amylase level falsely. How he obtained this knowledge is unknown; however this ruse is apparently known by some inmates in prison and used to their advantage. The patient's motivation was simple: repeated admission to the hospital for abdominal pain and hyperamylasuria enabled him to obtain narcotic analgesics for his apparent pancreatitis. We wonder how many other patients resort to such an ingenious, albeit fraudulent, practice. Of great interest was the observation that in three healthy subjects the addition of 2 ml of saliva to 30 ml of urine markedly elevated the amylase content of the urine. Points that should alert one to this phenomenon are an alkaline urine pH and urine containing excess epithelial cells, mucus, numerous white cells, and bacteria.

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EVOLUTION AND THE HUMAN TAIL

A Case Report

Fred D. Ledley, M.D.

THE birth of a child with a caudal appendage resembling a tail generates an unusual amount of interest, excitement, and anxiety. There is something seemingly unhuman about the presence on a human infant of a "tail" like the tails found on other primates. It is incongruous; it violates our sense of anthropocentricity, and it raises issues that involve not only teratology and embryology but also our view of ourselves and our place in evolution.

The human tail has long been an object of scientific curiosity.[1-3] During the early part of the 19th century the caudal appendage was considered to be an example of maternal impression. A report of a "child with a tail" as late as 1894 documented that the mother had held a baby pig by its tail during her pregnancy and concluded that the malformation in her offspring represented the maternal impression of this event on the fetus.[4] The caudal appendage achieved particular prominence during the debates over Darwin's theory of evolution. To evolutionists the "human tail" was an example of a "reversion to a lower species" and an illustration of the doctrine that "ontogeny recapitulates phylogeny."[2] Hundreds of cases of "human tails" (of various credibility) were reported between 1850 and 1900, during the heyday of recapitulationist theory and the height of the debates over Darwinism.[2,3,5-9]

As recapitulationist theory gave way to more modern concepts of ontogeny and phylogeny and evolutionary theory achieved a firm foothold in Western science and philosophy, interest in the caudal appendage waned. There have been only sporadic case reports during the latter part of this century. These reports emphasize the legendary and historical aspects and ignore the evolutionary implications of this malformation except as a historical artifact.[10-13] This report describes a case of a child with a well-formed caudal appendage and explores archaic and modern explanations for this malformation. The human tail serves as an example of modern concepts of ontogeny and phylogeny and presents a striking clinical confrontation with the reality of evolution.

Case Report

The infant was a 3400-g product of a normal full-term pregnancy. A caudal appendage was noted at birth, and the child was transferred to the Children's Hospital Medical Center in Boston. There was no history of maternal infection or exposure to drugs or known teratogens. There was no family history of any congenital anomalies. The appendage was 5.5 cm long and tapered at the tip, with a diameter of 0.7 cm at its base (Fig. 1). It was located 1.5 cm to the right of the midline adjacent to the sacrum. The appendage was

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covered by skin of normal texture and had a soft, fibrous consistency. The infant was otherwise entirely normal. Radiographs of the spine were normal. The appendage was removed under a local anesthetic on the second day of life and was found to have no connection to vertebral structures.

Histologically the appendage consisted of a fibrous, fatty core, with normal skin containing dermal and epidermal layers, hair follicles, and sensory neural patches. There were no bony or cartilaginous elements in the appendage.

Discussion

The isolated caudal appendage resembling a tail is a rare entity. This was the first case recorded in the files of the pathology department at the Children's Hospital Medical Center since at least 1936. It is a benign lesion that has never been reported to recur in families.[1] Clinically it is important only in that it must be distinguished from more serious lesions, such as spina bifida, caudal regression, or sacrococcygeal teratomas. Its notoriety derives entirely from its place in ancient legend and its interest as a model for the relation among human malformation, ontogeny, and phylogeny.

Numerous legends and scientific writings before the 19th century allude to the resemblance of embryonic, vestigial, and malformed structures of higher species to structures that are characteristic of more primitive species. In the mid-19th century, Haeckel provided a formal basis for these similarities in his famous aphorism that became known as the "biogenic law": "Ontogeny is the short and rapid recapitulation of phylogeny." [14] The process of evolution according to recapitulationist theory had two components: the "terminal addition" of developmental stages for higher organisms at the end of the complete embryonic development of lower organisms; and the acceleration, or condensation, of these sequential stages into the time course of gestation. It was believed that the fetus passed through the forms of lower species during the course of development. Many congenital malformations were thought to represent a developmental arrest at an intermediate stage of development and were regarded as reversions to the form of a lower species. In the recapitulationist model, embryology and teratology thus provided an important window on man's phylogenetic origins.

It was in this context that the caudal appendage captivated the attention of 19th-century science. For example, Darwin cites a report of human tails in *The Descent of Man*[15] and considers this anomaly in his reconstruction of human ancestry:

> By considering the embryological structure of man — the homologies which he presents with the lower animals — the rudiments which he retains — and the reversions to which he is liable, we can partly recall in imagination the former condition of our early progenitors; and can approximately place them in their proper place in the zoological series. We thus learn that man is descended from a hairy quadruped furnished with a tail. . . .[16]

When the caudal appendage is critically examined, however, it is evident that there are major morphologic differences between the caudal appendage and the tails

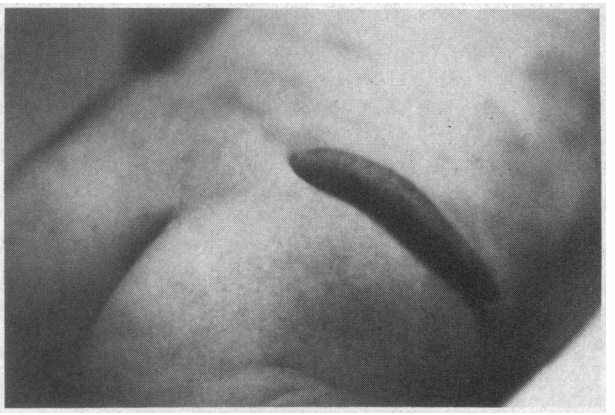

Figure 1. Photograph of the Caudal Appendage before Surgical Removal.

of other vertebrates. First of all, the caudal appendage does not contain even rudimentary vertebral structures. There are no well-documented cases of caudal appendages containing caudal vertebrae or an increased number of vertebrae in the medical literature, and there is no zoological precedent for a vertebral tail without caudal vertebrae (Jenkins FA Jr: personal communication). Secondly, the appendage is not located at the caudal terminus of the vertebral column. It is possible that this structure is merely a dermal appendage coincidentally located in the caudal region. This possibility cannot be excluded. There is a precedent for accessory digits on the hands and feet to contain various bony components, but dystrophic digits often contain no bony structures, although they may have other features, such as nails, that are characteristic of digits. In addition, mutant tails lacking vertebral structure or displaced from the midline are known in several tailed species, such as the mouse.

The human embryo has a tail complete with as many as 10 to 12 caudal vertebrae during the sixth week of gestation (14 to 16 mm).[17] At this stage the human embryonic tail is virtually indistinguishable from the embryonic tails of tailed species. During the seventh and eighth weeks of gestation the human tail regresses. There is a reduction in the number of caudal vertebrae by fusion, leaving the vestigial coccyx, and the projecting portion of the tail disappears as a result of the growth of other caudal structures.[9] Hughes and Freeman have compared the events of tail embryogenesis in vertebrates with tails and in those without tails, such as human beings and chickens.[18] They suggest that in species without tails the caudal extension of the neural tube ends, and the posterior neuropore closes, before formation of the tail bud. In tailed species the tail bud forms and is cannulated by the neural cord before closure of the posterior neuropore. Thus, in tailed species the neural tube and notochord extend the length of the embryonic tail, whereas in tailless species these elements are not present. In 1901 Harrison postulated that the human caudal appendage arose from the caudal filament repre-

senting the distal portion of the embryonic tail, which did not contain vertebrae.[9]

Many genetic variations of tail morphology have been studied in the laboratory mouse.[19] Tail length, with normal vertebrae, is associated with polygenic or multifactorial inheritance.[20] Over 30 specific genetic loci associated with mutations in tail morphology have been described. Most mutant tails in the mouse contain caudal vertebrae.[19] However, a mutant boneless tail resembling the human caudal appendage has been reported to be associated with the mutant allele *truncate*.[19,21]

The *truncate,* or *boneless,* phenotype in the mouse is produced by an autosomal recessive mutation with variable expression; in the homozygous state, it causes an absence of vertebrae in the caudal, sacral, or lumbar region.[19,21] Frequently, caudal vertebrae can be completely absent, leaving a shortened, boneless tail filament containing only loose connective tissue, blood vessels, and nerve fibers. The mutant tail is often displaced from the midline. The embryonic events leading to the boneless tail have been described in some detail.[21] The basic defect is that the notochord fails to extend properly into the tail of the developing embryo. Somite cells degenerate at levels where the notochord is absent. The embryonic tail thus contains no neurotube or neural cord, and it constricts, leaving only a filamentous appendage. The expression of the *truncate* genotype is variable. Some homozygous mice have no caudal vertebrae, and others have only segmental loss of vertebral structures.

What does comparative embryology teach us about the human caudal appendage? First of all, if I may be pardoned the anachronism, the human caudal appendage does not represent a regression to a lower species in the sense of recapitulationist theory. A devout recapitulationist might make the heuristic argument that the *truncate* mouse itself represents regression to a lower vertebral form. This absurd line of argument, often invoked during the heyday of the biogenic law, led to the creation of such mythical creatures as the moneron, blastaea, gastraea, and finally pithecanthropus to fit hypothetical evolutionary ancestors.[14]

Secondly, the structural elements involved in tail formation are almost identical in species with and without tails, in the *truncate* mouse, and presumably in the human caudal appendage. The morphology of the mature tail is determined by the temporal sequence and spatial relation among caudal structures at critical points during ontogeny. Thus, the timing of tail-bud formation in relation to closure of the posterior neuropore, and the spatial relation between the notochord and adjacent structures within the developing tail bud, determine the morphology of the mature tail. It is reasonable to postulate that a sequence of developmental events similar to those in the *truncate* mouse would result in a structure such as the human caudal appendage.

This model of the way in which the caudal appendage may have arisen reflects a modern understanding of ontogeny and phylogeny in general.[14,22] Differentiation is thought to be primarily related to variations in the temporal, spatial, or proportional relations between developmental structures and events. These variations, in turn, are thought to involve alterations in the timing and kinetics of gene expression that are secondary to mutations in regulatory genes. For example, there is an extraordinarily small difference between the structural elements of the human and the chimpanzee on a molecular level.[23] Immunologic, electrophoretic, and amino acid-sequence comparisons of homologous proteins from the two species demonstrate greater than 99 per cent identity. Studies of thermal stability comparing human and chimpanzee DNA demonstrate only slightly more variation in genomic structure. These studies suggest that the profound phenotypic differences between the human and the chimpanzee result from genetic changes in regulatory genes rather than from differences in structural genes.[23] The molecular nature of these mutations remains largely hypothetical.[22]

In modern theory the parallels between ontogeny and phylogeny derive from the ability to trace the phenotypic expression of developmental mutations to specific stages of embryonic development at which differentiation occurs between largely homologous molecular and morphologic structures. Teratology has an important place in this model. First of all, spontaneous genetic mutation is thought to be the driving force of evolution. Whether a specific mutation represents teratology or evolutionary change is simply a function of perspective. Secondly, some malformations may in fact represent back mutations to an ancestral state or examples of parallel evolution. Other similarities may result from random variation within the restricted phenotypic patterns available for the expression of similar structural elements.

The human caudal appendage thus serves as a model for modern theories of ontogeny and phylogeny, just as it served theories of maternal impression and recapitulation in previous centuries. The modern understanding of teratology and tail formation finds nothing unhuman or reversionary about this tail-like structure. Rather, the caudal appendage reminds us of the context and the continuity of human evolution. The child with a tail is striking not because the tail is a "reversion" but because it is *not* a reversion — because it is entirely consistent with our understanding of ontogeny and phylogeny, which places us in the midst of primate evolution. The occurrence of the caudal appendage, as well as the presence of a well-formed embryonic tail in a child, are testimony to the preservation of the structural elements necessary for tail formation in the human genome. Twenty-five million years have passed since human beings diverged from their most closely related tailed primates, and 5 million years have passed since the family Hominidae diverged from the family Pongidae (the apes)[24]; yet the genetic distance between us and chimpanzees is less than that seen among sibling species of other organisms.[23]

As the renewed visibility of the creationist movement indicates, these concepts of human evolution are disturbing to many people; they touch on the raw nerve of human anthropocentricity. Even those who are familiar with the literature that defined our place in nature — from Darwin's *The Descent of Man*[15] to Wilson's *On Human Nature*[25] — are rarely confronted with the relation between human beings and their primitive ancestors on a daily basis. The caudal appendage brings this reality to the fore and makes it tangible and inescapable.

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CASE RECORDS
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CASE 20-1982

Presentation of Case

A 42-year old woman was admitted to the hospital because of dyspnea and edema.

There was a history of ulcerative colitis of 19 years' duration. Several episodes of activity had been treated with adrenocorticosteroids and sulfasalazine. Two months before entry she began to have bouts of irregular cardiac rhythm that lasted 15 to 30 seconds. A physician found no abnormality on physical examination. An electrocardiogram revealed incomplete right-bundle-branch block. Periodic sigmoidoscopic examination disclosed evidence of mildly active colitis. The patient was advised to discontinue caffeine and felt improved. One month before admission she became fatigued and experienced dyspnea on exertion; these symptoms progressed slowly and were followed by the development of orthopnea and slight abdominal distention. She saw another physician, who made a diagnosis of bronchitis and prescribed erythromycin, without improvement in the symptoms. Anorexia developed, but her weight increased, the abdominal distention worsened, and peripheral edema appeared. She returned to her usual physician, who referred her to the hospital.

She had a hydatidiform mole 11 years earlier; vaginal examination with cytologic study was negative 11 months before entry. The patient smoked rarely and consumed little alcohol. There was no history of sweats, chills, cough, sputum production, arthralgia, rash, ocular or oral inflammation, chest pain, photosensitivity, Raynaud's phenomenon, pulmonary emboli, or tuberculosis. Twenty-five years earlier a girl friend was said to have pulmonary tuberculosis. The patient traveled to Puerto Rico and Bermuda in recent months without experiencing illness and received no medication during the two years before entry.

The temperature was 37.2°C, the pulse 60, and the respirations 18. The blood pressure was 120/80 mm Hg, without pulsus paradoxus.