The author has proposed in previous publications that isolated cardiac malformations have an evolutionary origin. This is partly supported by the fact that isolated cardiac malformations found in humans occur also in other placental mammals as well as in birds. External gross examination of the heart in just over 5,000 birds was carried out during a 3 year period. Anomalies included one instance of duplicate hearts, two specimens in which no heart could be identified and in a fourth, a yellow-rumped warbler, the heart lay in the neck outside of the thoracic cavity.

Published reports of similar occurrences of an ectopically placed heart concern birds, cattle and humans. The fact that various species of both placental mammals and birds show evidence of heritability for heart defects, and that these species cannot interbreed, combined with the fact that birds and mammals have many similar malformations, points to either a common external causative factor or a common origin.

Genes that code the malformed heart must be transmitted with that part of the genetic makeup common to all birds and mammals. Malformations caused by teratogens produce widespread organ injury to a potentially normal embryo whereas the evolutionary malformation is an organ-specific anomaly in an otherwise normal mammal or bird and occurs in widely separated species. The implications of this theory are important for parents of children with an isolated congenital heart defect who may have ingested one or another drug or chemical or have been exposed to toxins or infectious agents before or after conception of the affected offspring.

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FOREWORD

At the age of 86, Helen Taussig completed the major portion of this manuscript only a few days before her accidental death in May 1986. Several weeks before that date, she had commissioned some of her friends and former fellows to put her work in final form and to submit it for publication if “anything should happen” before she could do the job herself. After her death, this group met, read and reread the manuscript and agreed to edit it gently, retaining as much as possible of her unique expressions while striving for the brevity that she always aimed for.

Helen Taussig was a master at deductive reasoning. Witness the Blalock-Taussig operation. She was convinced that the procedure would work in patients, though no animal experimental model was available to test the proposal. Her concept of the evolutionary as opposed to the teratogenic origin of isolated cardiac malformations evolved from her talent for deductive reasoning. She had no intent to label this work a scientifically based research project. In fact, the editing group believed that it should be regarded as a treatise.

She had seen teratogens such as thalidomide and the rubella virus during pregnancy wreck multiple organ development in the fetus including, but not confined to, the heart. But concerning isolated cardiac anomalies, it never made sense to her that a teratogen could attack one or more components of the heart and leave other developing organs undisturbed. She had seen the distraught parents of a child with a heart defect, who were repeatedly asked by physicians about events during the pregnancy, blame themselves for whatever they might have done or consumed that allegedly led to the heart defect in their child.

This treatise addresses the very fundamental issue of the origin of isolated cardiac malformations. Whereas the observations in bird hearts do not provide scientific proof that these defects result from genetic transmission influenced by evolutionary changes in the genes rather than from embryonic exposure to teratogens, we cannot help but keep our
mind open (pending further investigations) to that possibility in our history taking and in counseling parents about their role in the occurrence of a heart defect in their offspring.

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Introduction

This study of cardiac malformations in Aves is a direct extension of my previous study (1) of common cardiac malformations that occur as isolated anomalies and occasionally in syndromes of multiple organ congenital anomalies in humans and many other placental mammals. I presented evidence that the origin of these malformations dated back to the evolution of the Mammalia from the Reptilia, approximately 100 million years ago.

Aves were chosen for this study because they belong to a different class in the animal kingdom and are of a similar antiquity. The avian heart closely resembles that of the placental mammals in that both have two atria, two ventricles, one main pulmonary artery and one aorta. An obvious anatomic difference is that in the placental mammals the aorta arches to the left and in the Aves to the right. Both are warm-blooded animals and, in both, the heart operates during fetal life as in the Reptilia, i.e., as a one-pressure system with admixture of venous and arterial blood. Within seconds after birth, in both birds and placental mammals, the circulation changes to a two-pressure system with virtually complete separation of arterial and venous blood.

Aves are believed to have evolved from the Reptilia through dinosaurs, a special group of Reptilia. The Mammalia also evolved from the Reptilia, but by a different, though unknown pathway. Therefore, I reasoned that if my theory of the evolutionary origins of the common cardiac malformations is true, birds should have a number of cardiac malformations similar to those of placental mammals and perhaps some different ones. Although the corollary to this is probably equally true, i.e., the placental mammals may have some malformations that birds do not have, this would be virtually impossible to prove. Therefore, I have limited my study to ask whether birds have some malformations similar to those of placental mammals and perhaps some different ones.

Review of the Literature

Duplicate hearts. The earliest report of a cardiac malformation in birds dates back to about 300 BC. Theophrastus (2) (67 to 297 BC) is reputed to have said that partridges in Paphlagonia sometimes have two hearts. This statement is repeated by Zlîny the Elder (3) (22 or 23 AD to 79 AD) and later by Aulus Gellius (4), Aelius Aristides (5) and Athenaeus (6). I found no further reference to duplicate hearts until the 18th century reports of Plantade (7) and D’Auberville (8).

Concerning duplicate hearts, Baillie (9), in a footnote, quotes Haller (10) as stating, “I personally possess two hearts from a goose, in which animal such an occurrence appears to be not rare.” In 1851, Meckel (11) also reported two hearts in a goose, and Panum (12) reported finding duplicate hearts in a chicken embryo.

In 1900, Constantinouzcu (13) wrote a beautifully illustrated report of a pigeon with two hearts. The hearts lay one on top of the other, with the ventral heart being slightly larger than the dorsal heart. Each heart had two atria and two ventricles. He gave the measurements of the atria and ventricles of each. Alas, the specimens had been torn out during cleaning of the bird so that it was impossible to determine how the circulation of the two hearts joined!

The most famous account of multiple hearts in birds is by Verocay (14): “It is a familiar fact that every now and then two hearts have been found in one hen, but surely the occurrence of seven hearts in one hen is worth recording.” Verocay, a pathologist, arrived at an inn in the Tyrol 5 minutes after the hen had been found to have seven hearts. He gave the date, name of the town, the name of the inn, the name of the innkeeper and that of the cook who found the hearts when cleaning the hen. Verocay said, “I would not have believed it if I had not seen it with my own eyes.” It took Verocay 2 years to persuade the innkeeper to give the specimen (which had been preserved in strong spirits) to him for his museum, the Deutsche Pathologische-Anatomische Institute in Prague. All hearts were of normal size and had two atria and two ventricles. The fact that all hearts were of normal size and none had atrophied indicated that they each had functioned. Verocay speculated whether all seven hearts beat simultaneously or in rapid succession. Inasmuch as the specimen had been found during cleaning, the circulation could not be traced (Fig. 1).

In 1951, Stanski (15) reported two hearts in a rooster that had been chosen for breeding because he seemed to be exceptionally strong and healthy but was later found to be too pugnacious for breeding.

In 1952, Mostafa (16) reported a case of four hearts in one hen of the Fayoumi breed. Each heart was of normal size and shape with two atria and two ventricles, and microscopy showed normal cardiac musculature.

Since I became interested in duplicate hearts, three of my friends have told me of finding duplicate hearts in domestic fowl—several in Plymouth Rocks and two in Rhode Island Reds.

*Paphlagonia is on the southern border of the Black Sea, a few miles inland from Sinope (ancient Sinope).
The question has been repeatedly asked whether duplicate hearts have ever been reported in humans or other placental mammals. Winslow (17) reported that Fontenelle, after a long search, learned that Collomb had demonstrated two hearts in a grossly deformed infant and reported this before the Royal Academy of Science in Paris. The monster had one cyclopean eye with one lens, two pupils and two retinas, no nose, no mouth, ears low-set at the level of the larynx and two hearts. Each heart lay in its pericardial sac, one pointing to the right, the other to the left, and shortly after arising from the heart, the two aortas joined to form a single arterial trunk. There was no duplication of any other organ. This is the only authentic report in the literature of two hearts in humans (14,18).

Ectopic hearts. In 1905, Waterston (19) reported an extraordinary ectopic heart from a wild duck, *Anas querquedula*, which had been shot in November 1904 in the Thano Jungles near Bombay. The bird had been sighted flying strongly and circling overhead for about five minutes before being shot down. It was only at the dinner table when a cut was made in the pectoral muscle that a cavity was seen. Thereupon the entire pectoral muscle was preserved in dilute alcohol and sent to Waterston for study. In the center of the muscle was a cavity three-fourths of an inch in length by one-half inch in width in which lay a perfectly formed avian heart. No pericardium was seen. The surface of the heart was covered by smooth epicardium. The heart was apparently normal and gave off from its anterior surface two great vessels that unfortunately had been cut too short to trace their course.

Tuttle and Greene (20) reported a 3 year old parakeet that had recently developed a soft fluctuant swelling in the episternal notch. The swelling slowly increased in size, therefore surgery was recommended. At operation, the structure was found to be a large hematoma apparently caused by the rupture of an epicardial capillary of an ectopic heart. It was thought that the parakeet had injured its neck flying against some sharp object in the wire cage.

Other Malformations in Birds

Atrial septal defect. In 1900, Borgherini reported three generations of pigeons with an atrial septal defect, presumably so severe that all but one pigeon of the second generation died shortly after being hatched. Autopsy of one of the pigeons showed an atrial septal defect 6 mm in diameter, an enormous defect for a young pigeon. It is worthy of note that Borgherini suspected a cardiac malformation by observing that the pigeon had rapid respirations not only when awake but also when asleep. He suggested that the heritability of cardiac disease in humans might be studied in animals.

Ventricular septal defect. Siller (22) studied the occurrence of ventricular septal defect in chicks that died during their growing period. The group was composed of 613 chicks from six different strains. He found 288 ventricular septal defects. Two were small defects of the muscular type and the remaining 286 were high ventricular septal defects lying beneath the aortic valve. Analysis of the data showed that 97% of the defects were in three strains of chicks and 84% were found in one inbred strain. Siller considered this strong evidence of heritability of the malformation. Many of the large ventricular septal defects showed "lips" or "crescentic folds" that grew out over the defect and decreased its effective size. In some instances the lips had grown together and closed the defect. Siller noted that this malformation was not known to occur in humans.

Siller and Hemsley (23) studied the incidence of cardiac malformations in 171,000 chickens in seven flocks of broiler chicks (these flocks were not inbred). During the 11 week growing period 5,444 chicks died or were killed. Siller personally examined these 5,444 chicks and found 31 (0.57%) with a cardiac malformation: 10 with ventricular
septal defect; 8 with atrial septal defect; 2 with combined atrial septal defect and ventricular septal defect; 7 with overriding aorta, 2 with double outlet right ventricle; 1 single ventricle and 1 hypoplastic aorta. Thus, 31 cardiac malformations in 5,444 chicks gave the incidence of 0.57%, which is remarkably close to 0.5% in dogs and 0.5 to 0.8% in humans.

In a third paper, Siller (24) reported the pathology of dextroposition of the aorta in chicks. He concluded it was similar to that reported by Patterson et al. (25) in their study of the embryonic heart of Keeshond puppies with tetralogy of Fallot. In seven of Siller’s chicks, the aorta overrode the ventricular septum, but none had tetralogy of Fallot.

In 1972, Einzig et al. (26) reported three instances of ventricular septal defect found during a study of round heart disease in turkeys. All were high ventricular septal defects and had “lips” similar to those described by Siller. In two turkeys, the ventricular septal defect was clearly visible, and in the third the lips had closed the ventricular septal defect.

Summary. This review of the literature clearly shows that birds do have a number of cardiac malformations of both the acyanotic and cyanotic types similar to those found in humans. In addition, birds have at least three malformations that have not been reported in any placental mammal: 1) “lips” that grow out from the sides of a large ventricular septal defect; 2) ectopic hearts lying externally on the sternum with no split in the sternum; and 3) duplicate or multiple hearts compatible with life. Duplicate hearts have been reported in partridges, geese, pigeons, domestic fowl—all birds that spend most of their lives on the ground. Hence, the question was raised whether duplicate hearts ever occur in birds of the wild. Therefore, I undertook a simple study of gross cardiac malformations in birds—malformations visible to the naked eye.†

Method of Study

Essential requirements. 1) The first essential requirement for the study of any protected birds, alive or dead, is to obtain a license or permit from the Fish and Wildlife Service of the state in which one works and from the Federal Fish and Wildlife Service, or to have access to an institution or individuals with such permits. 2) Any study of congenital cardiac malformations requires a large number of specimens. Annually, thousands of birds are killed by collision with radio and television towers and ceilometers (27). I have obtained my birds through the courtesy of Rob: E. Crawford, the ornithologist of Tall Timbers Research Station in Tallahassee, Florida, who collects the birds that are killed at the nearby WCTV tower, and from Charles M. Kemper, MD of Chippewa Falls, Wisconsin who collects birds that meet their death at the WEAU television tower in Eau Claire, Wisconsin. 3) Adequate deep freezer space in which to store the birds and an adequate disposal system are essential. 4) If one is not an ornithologist, the assistance of an ornithologist will be of great help in many ways: identification of birds (not labeled at source), their scientific names, correct order of tabulation, age and sex, help with the avian literature and many other unexpected ways.

Technique to expose the heart. 1) Pick up a bit of skin and feathers in the episternal notch and pull it back to the xiphoid. 2) Push back the skin and feathers on both sides of the sternum to expose as much of the ribs as possible. 3) Make an incision below the xiphoid and cut through the ribs parallel to the sternum. Place the cut at least halfway between the sternum and the vertebral column and extend the cut up to the clavicle. Do this on both sides. 4) Lift up the sternum at the xiphoid and turn it back over the clavicles. There lies the heart clearly visible. §

Examination of the heart. Identify the great vessels and trace them as far as you can. Next examine the posterior (dorsal) surface with care and try to discern the atra. It is important to become familiar with the posterior surface Constantinescu (13) described the two hearts in his pigeon as one lying on top of the other. The anterior ventricle was the one lying just beneath the sternum; the posterior or dorsal ventricle was slightly smaller and lay immediately behind the anterior ventricle. This I believe was the true relation of the two hearts and not as shown in Varocay’s illustrations in which the hearts are well separated and connected by strands of tissue (presumably pericardium). Search the abdomen carefully; if no heart is seen in the chest, the episternal notch and neck also should be examined with care for a misplaced heart.

Finally, excise the heart together with the great vessels starting on the anterior side and then turning the heart and excising the posterior vessels behind the heart, including as many of the pulmonary veins as possible, thereby trying to save the atrium. Put the hearts in separate jars. I have saved some of the hearts at the Delaware Museum of Natural History for future study by anyone interested.

Findings

Case material. I have examined slightly more than 5,000 birds during 2 academic years. The species of those birds are tabulated and available for reference at the Delaware Museum of Natural History in Greenville, Delaware. The vast majority (about two-thirds) of these birds were warblers from Wisconsin or Florida. Warblers are a subfamily, the Parulinae, of the family Emberizidae; they include many

†This seemed a suitable study for an octogenarian no longer working in a medical school or pathology laboratory and with no budget. I am deeply indebted to the Delaware Museum of Natural History for granting me the privilege of working there and assuming the cost of the study.

§This is an adaptation of Siller’s technique in chickens. He cuts a lap of skin at the xiphoid and yanks it back to the episternal notch and then pushes or pulls the skin back on both sides to expose the sternum. It is not necessary to pluck chickens or smaller birds.
different species that do not interbreed. Gross cardiac malformations visible to the naked eye are rare.

This study revealed three interesting findings in warblers of three different species, one from Florida and two from Wisconsin.

Duplicate heart. My first finding was in a yellow-rumped warbler from Tall Timbers, Florida. As I removed the heart I thought that I saw the tip of a second ventricle (not at all where I had expected to find a heart from the picture I had seen of Verocay’s heptacardia [14] and Mostafa’s four hearts [16]). I carefully removed the heart and put it in a separate jar to clear. The next day when I took the heart from the jar with my pincers, a second heart followed, but alas the strand between them was so thin that it broke before I could set down the specimen. The two hearts looked identical and were approximately of the same size (approximately 4 mm long and 3 mm wide). I cheerfully thought I would soon find another duplicate heart. This I never did.

Acardia? The second abnormality I found twice. The first time was in a chestnut-sided warbler. When I lifted the sternum and exposed the chest and abdomen, no heart and no vestige of a heart was found in the chest or abdomen! A year later, on examination of a bay-breasted warbler from Eau Claire, Wisconsin, when I lifted the sternum to expose the heart, again no heart was seen, and no vestige of a heart was found in the chest or abdomen or in the episternal notch or farther up in the neck. Circulation of the blood is essential for life! Did these two birds have some bizarre ectopic heart as in the case reported by Waterston (19), or did they have hearts of a different molecular structure that were less durable than the normal heart or did they have a true acardia?

Baillie (in 1794) (9) and Godgluck (in 1970) (28) both started their classifications of cardiac malformations with acardia and the duplicitas (or multiplicitas) cardia and ectopic cardia. Amorphous acardia is the common type. This condition occurs occasionally in twins—most frequently in cows (29) and rarely in humans. In this condition, one twin is normal and the other is an amorphous teratomatic mass of tissue with an umbilical cord extending to the placenta of the normal twin. It is thought that the amorphous twin gets its circulation from the pulsations of the normal twin’s heart. The only reference to acardia in humans that I have located is that by Daniel in 1776 (30). He reported a case of a house servant who had a spontaneous abortion at 7 months. The female fetus was severely underdeveloped and born dead. The head was well formed, but there was no trachea or esophagus and most of the internal organs were lacking. There was no heart. A large vessel lay close to the posterior abdominal wall, and from it a number of smaller vessels “fanned” out across the abdomen. The umbilicus was normal and the umbilical cord was attached to an apparently normal placenta. Daniel postulated that the circulation was maintained through pulsations transmitted from the placenta through the umbilical cord to the fetus.

This report together with the current studies in the use of intermittent high vibrations to improve ventilation in premature infants (and in newborn infants requiring cardiac surgery) made me wonder whether the intermittent rapid vibrations of the wings of small birds during flight could make it possible for birds with acardia and primitordial cardiac cells to survive. If so, this represents another malformation compatible with life in birds but not in humans.

Congenital translocation of the heart? The third remarkable finding was in a yellow-rumped warbler from Eau Claire, Wisconsin. Nothing unusual was seen on the external examination. As I started to remove the feathers at the episternal notch, I felt a mass and naturally thought it would be an ectopic heart. On removal of the sternum, no heart was seen. This chest looked similar to the chests of the two warblers in which I could find no heart. There was no evidence of an injury or of any hemorrhage: all of this was consistent with an ectopic heart. I immediately turned my attention to the mass in the episternal notch. To my surprise, the mass was composed of fatty material in which were imbedded the liver, some intestines and the stomach! Further exploration revealed the heart lying about 3 mm higher in the neck! Inasmuch as the rest of the bird appeared normal, I thought it was a most unusual malformation.

David 1.ilies (see acknowledgments) rightly raised the question whether the force of the bird’s collision with the tower had thrown the stomach and heart up into the neck. Donald Patterson (see acknowledgments) later examined the specimen and thought it was a congenital anomaly because there was no evidence of bleeding or any other injury. Therefore, I wrote to Robert Crawford (see acknowledgments) for his opinion, and with his permission I quote his answer in full:

My opinion as to whether the specimen you describe was rendered that way by violent collision or abnormal development can only be a subjective guess as I did not see the bird hit the tower, and have not even seen the specimen. However, I have examined thousands of birds from the WCTV tower (and other towers) and have never seen any translocation of organs as you describe. Injuries I have seen were, as near as I can remember, always of three types. First, the specimen would appear to be intact, but would have some brain hemorrhaging evident upon dissection. I had always assumed this to be evidence of a head-on collision. The second type was as the first, but the bird would have a broken wing or bill, and be largely intact except for the cranial injury. The third type, more unusual, would be severely mangled. This is most often the case with larger birds, and I had always assumed that these large birds struck a guy wire which tore them apart. The organs, however, always seemed to be in an appropriate place.

I leave it to each reader to decide whether the bird had a dislocation of the stomach and heart from a severe collision
or a congenital translocation of the stomach and heart, i.e., a most unusual congenital malformation.

Discussion

Incidence of cardiac malformations in birds. Our knowledge of cardiac malformations in birds is extremely meager compared with that in humans, in part because the subject has not seemed important and in part because information is difficult to obtain. Indeed, most of the early reports concern accidental findings of unusual anomalies. The few studies that have been done all have their limitations. Siller's study of ventricular septal defect (22) was limited to chicks that died during the growing period; thus, those that died at birth and those with a malformation compatible with longevity were excluded. McDonald's studies on mortality in wild birds (31-33) was undertaken to determine whether the deaths were caused by adverse environmental factors or communicable infections. No duplicate heart was found among the thousands examined.

My study was limited to small birds of the wild, the vast majority of whom met their death by collision with television towers. This means that all these birds had left the nest. Because most were killed during the autumn migration, they had lived for several months. Thus all the severe cardiac malformations incompatible with life never entered my study, which was limited to gross malformations visible to the naked eye. Nevertheless, the information available in the literature shows that birds have cardiac malformations similar to those in humans and other placental mammals, suggesting to me the evolutionary origin of the common cardiac malformations.

In brief, cardiac malformations in birds as well as in humans have been known for thousands of years. The incidence of cardiac malformations in placental mammals and birds is remarkably similar. Moreover, a number of the same malformations are found in birds and in placental mammals, including humans. For example, ectopic hearts occur in humans, cows, parakeets, and wild ducks; ventricular septal defect occurs in humans, cats, dogs, horses, domestic fowl, and turkeys; double outlet right ventricle defect occurs in humans, dogs, cats, and domestic fowl; and single ventricle occurs in humans, horse, antelope, and domestic fowl.

Role of heredity. Evidence that heredity is a factor but not the primary factor in the etiology of cardiac malformations in humans has been generally accepted. The breeding experiments by Patterson et al. in dogs (23) and Fox's experiment in rats (34) show that the same is true in other placental mammals. Borgherini's report (21) of three generations of pigeons with atrial septal defect and Siller's finding (22) of a high incidence of ventricular septal defect in one inbred strain of hens both indicate strong evidence of heritability of cardiac malformations in birds. In short, evidence of heritability in various species of placental mammals and birds is clear.

It is important to remember that only closely related species interbreed. Placental mammals and birds certainly do not interbreed! The fact that many species of placental mammals and birds show evidence of heritability and that these species cannot interbreed, combined with the fact that birds and placental mammals have many similar malformations, points either to a common etiology (external etiologic factor) or to a common origin (a common beginning).

Role of teratogens. In regard to a common etiology, teratogens certainly exist. The two classic examples of teratogens for humans are thalidomide and the rubella virus.

Birds lay eggs; therefore, once the egg is laid, nothing that the mother eats or does and nothing that passes into her circulation can affect the embryonic bird. Nevertheless, teratogens are known to injure embryonic birds in two ways. The first occurs if the nesting area is contaminated. The mother may eat the toxic substance and pass it into the egg yolks as they are formed. The second possibility is that a teratogen may be of such a nature that it may pass through the pores of the eggshell, as do oxygen and carbon dioxide, and thereby injure the unhatched bird. Two of the best known teratogens in birds and insects are DDT (a derivative of dichlorodiphenyltrichloroethane) and PCB (a polychlorinated biphenyl compound).

Gilberston et al. (35) studied abnormal chicks in a nesting area on the lower Great Lakes. They found that many young terns were severely injured—their skeletons were weak and their feathers were reduced in number and so poorly formed that many of the young birds could not fly and consequently died. The eggshells of these terns contained varying but relatively small amounts of PCB. In the same nesting area, Gilberston et al. (35) also found PCB in far greater amounts in the eggshells of young herons, and these young herons were uninjured. They interpreted this observation as indicative of a difference in species susceptibility to the teratogenic agent, which is strong evidence that teratogens affect birds and mammals in the same way; i.e., a teratogen causes widespread injury to a susceptible species but does not injure all species.

Thus, it seems highly improbable (except for the forces of evolution that affect all life all the time) that a teratogen or a combination of teratogens exists that affects all birds and placental mammals in a highly specific way at a particular time; their embryonic development and, moreover, that has been operative throughout the world for hundreds and probably for many thousands of years. In contrast to the great improbability that a common external causative agent will ever be found for the cardiac malformations in placental mammals and birds, the similarities may result from their having common phylogenetic origins.
Genetic considerations: role of evolutionary origin of cardiac malformations. One of the basic concepts of evolution is that the Aves and the Mammalia evolved from the Reptilia. By the very nature of this concept, the avian and mammalian hearts evolved from the reptilian heart. It is reasonable to believe that the cardiac malformations that occur in both the Aves and the Mammalia came into existence or were in existence during the period when the "normal" heart of humans was evolving. It follows that the genes that code these "malformed" hearts must lie deep in that part of our genetic makeup that is common to all birds and placental mammals.

I am not a geneticist, nor am I an authority on evolution. It is, however, easier for me to conceive of the origin of cardiac malformations in broad terms of evolution than in precise genetic terms. Where do all our genes come from if not through evolution? Some day I believe evolution and genetics will come together, as indeed they must.

Therefore, let me give some evidence in favor of an evolutionary origin of cardiac malformations based on three fundamental concepts of evolution: 1) Evolution occurs through the chance mating of many millions and billions of recombinations and mutations of the DNA molecule. 2) The survival of the fittest. 3) Marked changes in evolution may have occurred through sudden and abrupt catastrophic changes in the earth and its atmosphere. The evolution of the Aves and the Mammalia from the Reptilia represents a great change, probably the greatest single step in the evolution of animal life. It marks the change from poikilothermal animals to hematothermal animals and a corresponding change in their entire metabolism and their hearts and circulation.

The heart of all poikilothermal animals operates as a one-pressure system in which the arterial and venous blood is not completely separated. Whereas the heart of a hematothermal animal operates as does the heart of a poikilothermal animal during fetal life, within seconds after birth the hematothermal circulation changes to a two-pressure system with complete separation of arterial and venous blood. Two circulations are thereby established with complete separation of arterial and venous blood. This is indeed a great step in evolution and may well have been precipitated by some great catastrophic change in the earth, its atmosphere and its temperature.

Evolutionary types of cardiac malformations. During the evolution of the avian and mammalian hearts from the reptilian heart many types of hearts must have formed. The normal avian heart is remarkably similar to that of placental mammals with the exception of the right atrioventricular valve. This valve in placental mammals has three leaflets, whereas in birds the valve, similar to that of the crocodile, is a muscular ring that contracts and relaxes.

The survival of the fittest does not mean that only the fittest survive. A number of other hearts, although not able to support life as long as our normal heart, continued to evolve and are able to support life for varying periods of time. These hearts we call "cardiac malformations." Some of these hearts do not completely separate the venous blood from the arterial blood; these we call "malformations causing persistent cyanosis."

We also see malformations that represent stages in the evolution of our normal heart. For example, an atrioventricular communis (an endocardial cushion defect), a persistent ostium primum, an ostium secundum defect (atrial septal defect) and a high ventricular septal defect all represent stages in the development of the normal heart. In addition, there are three types of hearts that are normal in poikilothermal animals that arc prototypes of cardiac malformations in hematothermal animals; to wit, the normal bicaudate heart of a fish occurs in humans as a rare malformation seldom compatible with life for more than a short time and the biventricular heart of a frog is familiar to us as a single ventricle. The normal four-chambered heart of the crocodile, which is always deficient at its base, is the prototype of the high ventricular septal defect so common in placental mammals and birds. The more primitive the prototype, the less compatible is the malformation with the survival of hematothermal animals. Primitive prototypes, however, do indicate stages in the evolution of our normal heart and show that the genes that code the malformations came into existence before the evolution of the Aves and the Mammalia from the Reptilia; they are on the down slope of evolution.

I do not mean to imply that these are the only cardiac malformations that are evolutionary. I propose that all of the malformations that humans have in common with other placental mammals or with birds, or both, are evolutionary in origin. Furthermore, there is every reason to believe that birds have many more malformations in common with placental mammals than have yet been found.

Let me clarify the essential differences between congenital malformations caused by teratogens and those of an evolutionary origin. A teratogen causes widespread injury to a potentially normal embryo or fetus of a specific species. An evolutionary "malformation" is an organ-specific abnormality; i.e., it affects one organ in a particular way in an otherwise normal placental mammal or bird and occurs in widely separated species. Evolutionary malformations are, I believe, genetic variants.

Conclusions. In brief, this study shows that birds do have a number of cardiac malformations similar to those in humans and other placental mammals—a finding that is consistent with their common origin from the Reptilia. In addition, birds have some malformations not known to occur in placental animals. This finding is consistent with the evolution of these two classes of animals by different pathways—the Aves from dinosaurs and the Mammalia by a different but unknown pathway.

All of these facts indicate that the genes that code the common cardiac malformations that occur in otherwise
"normal" individuals, other placental mammals and birds came into existence or were in existence during the evolution of the Mammalia and the Aves from the Reptilia. Hence, these malformations must be genetic in origin.

Indeed, if this theory is correct, it is logical to believe that the majority of other isolated congenital abnormalities are also evolutionary in origin. This means that neither exposure to toxic substances nor the parents can be held accountable for the occurrence of congenital abnormalities because such malformations represent just a few of the many manifestations of the evolution of life and its infinite variety.

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Working at the Delaware Museum of Natural History has been a happy experience. My thanks to all of the many mentioned persons and also to my many friends who have contributed the "miscellaneous" birds and have helped me in various other ways.

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