Congenital Malformations of the Heart

Embryology, Anatomy, and Operative Considerations

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Over the years, we often have been asked why successful open-heart surgery in man originated at the University of Minnesota when, outwardly at least, several other centers seemed better prepared. The answer always has been very clear to those of us who were there at the time. Namely, it was the presence of Owen H. Wangensteen, a truly visionary surgeon, as Chairman of the Department of Surgery. His lifelong recognition of the relevance of basic science and of the insight to be derived from research in the training of young surgeons created the milieu and opportunities for great achievements by many of his pupils, some of whose talents were not immediately obvious to others. John Hunter's immortal dictum to Jenner, "Don't think, do the experiment," was long ago changed by Owen Wangensteen to: "*Do* the experiment! And *do* think!"

Foreword

Accomplishment has been and continues to be the hallmark of professional and public recognition and acceptance. For centuries, the surgeon who was unlettered and trained only by apprenticeship was scorned by medical faculties. In 1536, the barber–surgeon Ambroise Paré substituted, with great success, turpentine, oil of roses, and egg yolk for Giovanni de Vigo's "oyle of Elders scalding hot" in the treatment of battle wounds at Turin. He thereby initiated the first systematic installation of an antiseptic into contaminated wounds, an occurrence that gained royal favor for surgery from a succession of French kings. When Charles-François Félix cured Louis XIV's fistula-in-ano by operation in 1686, surgery in France achieved a spirited public awakening, the appreciation, and the royal favor that lasted for more than half a century. It lead to the creation of five chairs of surgery at St. Comé under Louis XV (1724) and at the *Acadêmie royale de chirurgie* (1731). These were easily achieved despite vigorous protests from the Paris Medical Faculty.

In 1747, the British naval surgeon James Lind took twelve sailors who were ill with scurvy to sea on a brief Mediterranean cruise. Each was restricted to a basic diet of gruel with sugar for breakfast, fresh mutton broth for dinner, and barley, raisins, currants, sago, and wine for supper, with pudding and biscuits at other times. Lind divided his twelve patients into six groups. Each group was given a special increment to its basic food allowance. In six days, the two sailors who received two oranges and a lemon each day were already seaworthy and free of symptoms. It is undoubtedly one of the most critical, carefully controlled, telling, yet simple experiments ever performed in the history of medicine, with important consequences for and at minimal cost to seafaring military medicine.

In 1798, Edward Jenner contracted cowpox. Schooled in scientific inquiry by John Hunter, he was able to convert a country tradition of milkmaids' immunity to smallpox to a scientific reality, truly one of medicine's finest gifts to mankind.

Two of surgery's greatest innovations came from long-neglected Cinderellas of the profession: Boston's dentist W. T. G. Mortan brought anesthesia to surgery in 1846. In 1847, the Hungarian-born obstetrician Ignaz Semmelweis advocated prophylactic surgical antisepsis. This latter development has constituted, since the waning years of the nineteenth century, the seminal influence and very core of successful surgery. It superseded in effectiveness the introduction of antiseptics into wounds, the method long favored by surgery's first scientifically trained surgeon and great public benefactor, Lord Lister. Antibiotics, one of greater medicine's most precious gifts to mankind and a product of the twentieth century, has now joined these two most significant achievements for surgical progress in the nineteenth century.

In 1853 at age 40, Claude Bernard, surgeon-turned-physiologist, emphasized for all of the medical world through his clarity of thinking, intuitive reasoning, and critical experiments the need for a spirit of scientific outlook and inquiry that would demand testing of the validity of popular and prevailing theses by critical experiments. This doctrine provided a durable challenge to the profession. It was skillfully sketched by Bernard in 1865 in his *Introduction to the Study of Experimental Medicine.*

Today, a few leaders among American surgical academicians have characterized surgical research as research performed by surgeons. This is an extremely myopic outlook for surgery's advance.

Certainly open cardiac surgery is the discipline's most admirable contribution to greater medicine in this century. When medical school deans and search committees sought chairmen for their departments of surgery four to five decades ago, they were lucky to find a young surgeon who professed an interest in research. There were few candidates who could provide evidence of active and productive participation in scientific inquiry. During a *Wanderjahr* in Europe, this surgeon stood in awe and wonder one day in May 1928 before the Kaiser Wilhelm II Institute of Research at Dahlheim on the periphery of Berlin like a penniless youngster admiring the many delicacies on the shelves of a bakeshop, painfully aware that wishes do not have wings. World War II and the largesse of the federal government, urged on by Vannevar Bush, Franklin Roosevelt, Lister Hill, John Fogarty, and a sympathetic Congress, changed all this. Under the able supervision of James Shannon at the National Institutes of Health, there gradually emerged in our great universities a heightened, vigorous, buoyant, and sustained interest in research such as had never before been observed in our country. The transformation, new posture, and heightened international status of American medicine and surgery, attending the beginnings of systematic contributions to the patrimony of medical knowledge, challenge our credulity. The many accomplishments defy our understanding. During the first three decades of this century, the United States could boast only two Nobel laureates, both foreign-born: Alexis Carrell (1912) and Karl Landsteiner (1930). Today our biological scientists compete favorably in numbers and quality with the finest in the world, solely through the wonderful catalyst of research. May the public's belief and confidence in its miracles never cease!

Robert Gross of Harvard spearheaded the approach to cardiac surgery by his significant contribution to successful ligation of patent ductus arteriosus (1939) and division (1944). This was followed by the demonstration that coarctation of the beginning of the descending aorta could be dealt with satisfactorily by direct surgical attack, an achievement he shared in 1945 with Clarence Crafoord of Stockholm. Also in 1945, Alfred Blalock and Helen Taussig of Johns Hopkins startled many medical and lay audiences by their revealing photographs of deeply cyanotic children. The cheeks, lips, and fingers of these children had assumed a lively pink color attending increased oxygenation accompanying attachment of the proximal end of the divided subclavian artery to the pulmonary artery. Thus, an opportunity for improved oxygenation of the blood was provided by its second chance for passage through the lungs.

The Philadelphian John P. Gibbon, Jr., working in the laboratories of the Massachusetts General Hospital, lent an energetic propelling force to open-heart surgery. He demonstrated in 1939 that a pump oxygenator could sustain the canine circulation attending ligature of the pulmonary artery. The world's first successful "dry" open heart operation was performed in the University of Minnesota's surgical clinic by F. John Lewis on September 2, 1952. It was achieved by occluding both the superior and inferior vena cava for seven minutes for a patent atrial secundum septal defect under systemic hypothermia. This procedure materially reduced the oxygen requirements of the vital cerebral centers. On May 6, 1953, Gibbon successfully duplicated this performance, employing a pump oxygenator on an atrial secundum defect.

Less than a year later, on March 26, 1954, C. Walton Lillehei and his associates Richard L. Varco, Herbert E. Warden, and Morley Cohen successfully closed a ventricular septal defect in a 13-month-old infant. This was a far more complicated congenital cardiac defect. Within another year, this Minnesota team had achieved an astonishing record of other intracardiac surgical firsts, including correction of infundibular pulmonic stenosis, repair of atrioventricular communis defects, and successful anatomic correction of tetralogy of Fallot. Dr. Lillehei was joined by Dr. Richard De Wall, a newcomer to the cardiac team, and by March 1955, they had perfected for human use a simple disposable bubble oxygenator. This oxygenator is still used by cardiac surgeons all over the world. The further development of the self-contained unitized plastic bag oxygenator in 1956, with De Wall and Dr. Vincent L. Gott, lent an explosive impetus to intracardiac surgery. Successful operations for repair of acquired valvular heart disease followed quickly. On January 30, 1957, Lillehei's revolutionary concept for the control of complete heart block, with an electrical pacemaker connected directly to a myocardial electrode, appeared.

Many cardiologists and surgeons were amazed by these startling achievements of the Minnesota surgeons. How did it come about? This writer's contribution to the overall achievement was minimal: I was simply striving to create an atmosphere friendly to learning and scientific inquiry. I had been fortunate in gaining the opportunity to guide the destinies of the University's Department of Surgery in January 1930; I ill deserved it, but two far more likely candidates had rejected the opportunity saying, "There is nothing here and never will be." They had overlooked the burgeoning climate of research emanating from the preclinical departments of the University, a spirit that President George Edgar Vincent had brought to the Medical School in 1911 through the simple expedient of the power of appointment. Dean E. P. Lyon, from whom this writer acquired his opportunity, had said that the greatest dean he knew was Kipling's Gunga Din, regimental water carrier, an observation this surgeon took to heart with some effort directed to sideline cheerleading. Perhaps my only real participation in the endeavor was realization of the compelling need for surgeons to acquire an intimate knowledge of the physiology of the circulation. When my great friend and colleague, Maurice Visscher, returned to the University in 1936, I persuaded him and his staff to join us in a weekly surgical physiologic conference in a lecture amphitheatre at the hospital. These meetings continued until my retirement on June 30, 1967. Out of those weekly sessions came a number of extremely useful suggestions for surgeons. Perhaps the most fruitful of all the advantages was that a succession of brilliant and talented young surgeons—including not only Varco, Lillehei, De Wall, and Gott, but also Clarence Dennis, K. A. Merendino, Gilbert Campbell, Richard C. Lillehei, Castaneda, Morris Levy, Norman Shumway, Christiaan Barnard, and a number of others—spent two to three years or more fulltime under the tutelage of Visscher and his component physiology staff. Visscher has been generous enough to say that the arrangement was by no means of unilateral advantage only to the department of surgery.

Lillehei was the leader of this distinguished band of cardiac surgeons. He began his work in the University's Department of Surgery on October 24, 1945. He was still in uniform as he had just returned to the United States as a much decorated army veteran of 39 months overseas, and he was not receiving a stipend. By January 1, 1946, I had found shoestring support, a thesis which my mentors Dean

Lyon and President Lotus Coffman ardently supported. There was in fact no regular vacancy in the department nor available money for support of a new position, but my staff had become accustomed to moving over a bit to share opportunity with a promising new appointee. Sharing had become a byword in the department. I realized that men of promise were more difficult to find than money. Lillehei's first year's appointment carried an annual stipend of $1098. It was all the support money I could find. It paid off handsomely, not only for the University's medical school but for the world of medicine. Within four years, Lillehei rose to an associate professorship, the most rapid progression in the annals of our department.

This volume is a broadly based mature effort and a masterful presentation by its two erstwhile Minnesota surgeons. *Congenital Malformations of the Heart: Embryology, Anatomy, and Operative Considerations* should find warm acceptance and appreciation among cardiologists, physicians, and surgeons who are anxious to clarify their knowledge of and skill in the management of a host of congenital defects. Some of these are described and explained here for the first time.

Owen H. Wangensteen, Ph.D., M.D.

Preface

"There are in fact four very significant stumbling blocks in the way of grasping the truth which hinder every man however learned, and scarcely allow anyone to win a clear title to wisdom; namely, The Example of Weak and Unworthy Authority, Long Standing Custom, the Feeling of the Ignorant Crowd, and the Hiding of our Own Ignorance While Making a Display of our Apparent Knowledge.

"Every man is involved in these things, every rank is affected. For every person, in whatever walk of life both in application to study and in all forms of occupation, arrives at the same conclusion by the three worst arguments. Namely; This is a Pattern Set by our Elders, This is the Custom, This is the Popular Belief: Therefore, it should be held."

Roger Bacon on the Causes of Error
***Opus Majus*, thirteenth century**

The 1940s were an exciting decade in the progress of cardiac surgery. Up until that time, it had lain dormant for so many years.

In 1939, however, Dr. Robert Gross triggered this modern era with his ligation of a patent ductus arteriosus. The well-known contributions and successes of Gross, Blalock and Taussig, Crafoord, Bailey, Harken, Sellors, Brock, and others were the highlights of the rest of these years and dazzled the imagination of this author during his residency training.

The success of these early operations in and around the heart encouraged a handful of bold surgeons to begin thinking seriously of operations within the open heart. At that time, this still presented a seemingly impenetrable anatomic barrier to truly incredible possibilities. Although by the early 1950s, Dr. John Gibbon, Jr., had reported his history-making experiments with cardiopulmonary bypass, and intensive research on the development of pump oxygenators was vigorously pursued in a number of academic centers, by 1952 the surgical world had become thoroughly discouraged about and disillusioned with the clinical feasibility of open-heart surgery. By this time, many of the most experienced investigators had concluded, with seemingly good logic, that the problems were not with the heart-lung machines. They believed that the sick human heart was exhausted by failure. It could not possibly be expected to tolerate the cardiotomies, internal incisions, and stitching that were so successful when these same machines were used in healthy experimental animals.

Up until that time, a dozen or more open-heart procedures had been attempted in man in various parts of the world with extracorporeal circulation. Immediate failures had been uniform. There was one successful operation in May of 1953, when Dr. John Gibbon closed an atrial secundum defect in a young adult. Although this success was well received, it did not arouse much excitement among cardiologists and cardiac surgeons. This was because Gibbon was never able to repeat his one success with this simple defect or to achieve any success with the more complex ventricular defect. After four unsuccessful operations in 1953, he became discouraged and did not use his machine again. Moreover, Dr. F. John Lewis, a colleague at the University of Minnesota, was already at that time closing atrial septal defects under direct vision utilizing simple hypothermia. He had been doing this routinely two to three times a week with a high degree of success dating from his first successful operation with that technique on September 2, 1952.

Thus, the demonstration in early 1954 by the University of Minnesota surgeons that severely congenitally malformed hearts could be successfully repaired and made to function normally by utilizing a temporary period of extracorporeal perfusion to achieve an empty open heart understandably startled the medical world. Further, it was shown that these operations for complex anomalies could be performed repeatedly with predictable results and a surprisingly acceptable risk for critically ill infants with conditions heretofore considered hopelessly uncorrectable.

Initially, the extracorporeal oxygenation was achieved by cross-circulation of the blood between the patient and his parent. The cross-circulation technique was as strikingly successful in humans as it had been in the hundreds of dogs in the experimental laboratory. Although the clinical use of cross-circulation was an immense departure from the established surgical thinking and practice of the preceding 20 years, we had viewed it as a near perfect perfusion system since it simulated a temporary "artificial placenta." This fact was undoubtedly very important to the seemingly incredible accomplishments that were to follow. Within five months, a rapid succession of surgical firsts occurred (as already enumerated in the Foreword). Forty-five cross-circulation operations, all for complex congenital abnormalities once considered uncorrectable and therefore inoperable, were performed during the first 12 months.

The unprecedented success of the cross-circulation technique in patients with complex defects, and often in intractable heart failure, played a crucial role in rapidly dispelling the widespread pessimism that prevailed at the time among cardiologists and surgeons concerning the feasibility of open-heart surgery in man. The conclusion that the sick human heart represented an insurmountable barrier was clearly untenable, and attention was directed once again to the previously unrecognized deficiencies of the existing pump oxygenators and to perfusion physiology. The revelation that safe perfusion of the body could be maintained with several lengths of plastic tubing, a few clamps, and a little oxygen had an explosive effect upon the development of cardiac surgery. The surgeon's dream of performing intracardiac corrections in the open heart had become reality.

However, the phenomenal rapidity with which open-heart surgery developed following the widespread availability of this simple, efficient, and inexpensive equipment for perfusion predictably left large and significant gaps in our knowledge in all phases of this burgeoning new field of surgery: diagnosis, perfusion physiology, surgical anatomy, reparative techniques, and postoperative care, to mention only a few. Among these many unknowns, perhaps the most acute was the relentless pressure on the surgeon to recognize, through a limited operative incision, the seemingly endless variety of congenital malformations.

More than any other field of surgery before or since, successful open-heart surgery placed a high premium on efficient teamwork. The heart remains the sole circulatory organ, and the patient paid a severe price for inaccurate diagnoses, the operators' faulty or incomplete recognition of the functional pathology, and technical errors (be they valvular insufficiency due to the misplacement of a single stitch, hemiparesis due to a few cc of air or clot, or a lethal arrhythmia from poorly placed stitches in the Bundle of His). No amount of teamwork could offset such misadventures. This obvious strain on the surgeon's capability and performance was profound. It was exacerbated by an almost complete lack of accurate anatomical and pathologic descriptions of these lesions from the operative viewpoint, despite the fact that some lesions had been recognized for hundreds of years. In those early days of open-heart surgery, the only publications at all helpful were the truly remarkable monograph of Rokitansky, *Die Defecte Scheidewande Des Herzens,* published in 1875, and the *Atlas of Maude Abbott,* published in 1936. But these and other works had been undertaken at a period before operative treatment had even been dreamed of by their authors, and consequently they were insufficiently detailed for the surgeon's use. Ventricular septal defects, for example, were invariably pictured through a widely open left ventricle, and the vast spectra of the tetralogy or atrioventriculoris communis variations were scarcely noted.

Even more disheartening to the pioneer clinical surgeon were trips to the Department of Pathology to examine their collections of specimens. The state of affairs observed after death in muscular contraction aggravated by formalin fixation with the tissues frozen to the consistency of leather was so discouraging that our innate confidence that these lesions could be corrected was severely shaken. Fortunately, we instituted a plan whereby the pathology room diener would call us, day or night, for every autopsy performed on a baby or child, regardless of the diagnosis. Over a period of several years antedating the availability of any method for open-heart surgery, we had, through the cooperation of the pathology residents, the opportunity to open the hearts of a dozen or more patients with various congenital defects via a surgical cardiotomy and to repair the malformations. One of the most exciting memories of that period was the night we discovered how relatively easy it was to completely correct the intracardiac pathology in an infant who had died of the tetralogy malformations. Many times in subsequent years, we were to be poignantly reminded how incomplete was our knowledge of functional pathology, but working at that time with soft and pliable tissues restored our confidence that almost any defect within the heart could be corrected if the surgeon fully recognized the pathology and had sufficient time in an open bloodless heart.

None of the pioneer surgical teams were immune from these constantly humbling experiences of suddenly facing unfamiliar puzzling or even unknown pathology. Thus, there has been a relentless surge to rapidly secure solutions to these problems. While our judgment must be conjectural, it is the opinion of these authors that the cardiac surgeons of this past generation, highly competitive by nature and steadfastly independent by training, have every reason to be proud of the rapidity with which these vast problems have been overcome. Never before in medical history has there been such widespread spontaneous cooperation in the sharing of experiences, ideas, and methodology. This fact, as much as any other, has been responsible for the astonishing rapidity with which this exciting, new, and major field of medical treatment, open-heart surgery, has developed. Today, scores of pathologic conditions, previously hopeless and in some instances unrecognized, are being managed successfully in a routine manner even in neonates by surgical and medical techniques developed within this brief span of time.

Thus, it is all the more remarkable that in this same generation which has seen such an explosive proliferation in the quantity of medical literature, there has been no attempt to present a comprehensive description from the operative viewpoint of the pathologic anatomy of congenital heart disease in its wide spectra of variations, from the simple to the complex. There have been, of course, countless excellent papers, many symposia proceedings, and several superb monographs on various facets of the field that have vastly expanded our knowledge; but no one has attempted to put it all together.

The authors, having made such an attempt, can well appreciate the reasons for the lack of such a definitive work. It has proved to be a very formidable undertaking. This account of congenital heart disease from the surgical viewpoint was first completed in 1967 on the basis of our operative experience at that time in 2500 open-heart procedures. However, its publication was deliberately delayed because of certain misgivings about the rapidity with which operative techniques were still evolving and further apprehensions about the depth of our embryologic knowledge, so essential to the understanding of the pathologic patterns and their innumerable variants within these hearts. It seemed at that time that even after 15 years of astonishing progress, open cardiac surgery refused to lie still long enough for a definitive examination. While these delays have been frustrating personally, it is our belief that a better publication has been the dividend.

This volume is based on the authors' experiences in the fields of embryology and pathology and on observations made during operative repair of these malformations stretching over more than 20 years. The study of the embryology was made on microsections of embryonic human hearts in the Department of Embryology at the Carnegie Institute of Washington. During the course of these extensive embryological studies, several original observations were made which have direct significance for the clinician. In addition, the authors, aided by the personal knowledge gained in these embryologic studies that were augmented by the experience in the operative theatre, have systematically examined and studied in detail hundreds of specimens in the Department of Pathology at the University of Minnesota Medical Center, in the laboratory of Dr. Jesse Edwards, and in the Department of Pathology of the New York Hospital-Cornell Medical Center.

During these combined embryologic, clinical, operative, and autopsy studies, some significant original observations and concepts have evolved in the understanding of the developmental anatomy and classifications of complex lesions. We refer particularly to our observations on the anatomy of the atrioventricularis communis malformations, the developmental anatomy of the ventricular septum with relevance to the position of ventricular septal defects, the dextroposition complexes (including tetralogy of Fallot), the transposition complexes (including the various types of double outlet ventricles), and the embryology of the conotruncus and the ventricles. The probable embryological basis for the puzzling aortic-left ventricular tunnel has also been described. Some of these original observations have been published previously in journals, while others are published here for the first time.

We have chosen in our operative considerations to emphasize the preferred method of repair for the particular lesion being considered. This is a necessity in a single volume that deals extensively with developmental anatomy, pathology, and surgical correction of such a broad subject as congenital heart malformations. Where there is no consensus of opinion, we have attempted to evaluate the various procedures available. Likewise we have attempted, in discussing detailed operative approaches, to emphasize the important points of technique as well as the pertinent complications to be avoided. We have purposely avoided a step-by-step description of each repair as we felt that it was unwieldy and unnecessary in a volume of this type since there are several excellent monographs and atlases with the "how to do it format" available. In addition, we have included a rather extensive bibliography of the original as well as the significant subsequent contributions to which the reader may turn for additional information given in greater detail.

It is our earnest hope that this volume will be of significant value to cardiac surgeons (experienced as well as in training), cardiologists, radiologists, pathologists, embryologists, and, perhaps most important of all, to our patients—many as yet unborn.

C. Walton Lillehei, Ph.D., M.D.
Daniel A. Goor, M.D.

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The Anatomy of the Heart

INTRODUCTION

Congenital malformations of the heart tend to involve developmental segments, and, in order to understand them properly, one must be familiar with the embryology and, particularly, with the developmental anatomy of the heart. Therefore, this chapter covers the gross anatomy of the cardiac chambers with particular emphasis on their developmental nature. Anatomical structures are often defined in the light of pathologic conditions because this is eventually what we deal with.

DEFINITION OF TERMS

Determination of Type of Chamber[20,22]

The four chambers of the heart are named by their specific anatomic nature. For example, the right ventricle is termed *right ventricle* even if it is located on the left side of the septum. The specific features of each chamber are primarily related to the septal structures.[22]

Right atrium. The right atrium is identified by the limbus of the fossa ovalis. Normally, additional structures—such as the orifices of the vena cava, the coronary sinus, and the crista terminalis—are present in the right atrium. In pathologic conditions, however, one or more of these structures may either be absent or enter the left atrium. For this reason the atrial septum, even if only partially present, is the most reliable structure for identifying the right atrium.

Left atrium. This chamber is identified by septal remnants of the septum primum.

Right ventricle. This chamber reveals heavier trabeculations than the left ventricle. In addition, heavy muscular bands traverse the right ventricle from septum to parietal wall. The term *moderator band*[18] has been utilized for the stoutest of these bands.

Whenever the ventricular septum is present, the chordae of the septal tricuspid leaflet are inserted into the septum. According to some authors, a *crista supraventricularis* (conus septum) is present in the right ventricle.[20,22] This is correct for the normal right ventricle. In some pathologic conditions, however, the crista supraventricularis may be absent.

Anatomic left ventricle. The trabeculations of this chamber are finer than those of the right ventricle. Whenever a septum is present, its base is devoid of trabeculations and there are no chordal insertions from the mitral valve into the septum.

Atrial Situs

Atrial situs refers to the relative side position of the atria in the chest. That is, whether the anatomic right atrium is on the left or right side of the anatomical left atrium. The right atrium on the right side is known as *situs solitus* of the atria; the right atrium on the left side is called *situs inversus* of the atria.

Since in certain conditions the atria are in situs solitus but the ventricles exchange sides, the atrial situs is not necessarily indicative of the remainder of the heart.

When cardiac malformations are not present, the general term situs solitus implies a normal situs of the atria and a normal heart. Similarly, the general term situs inversus indicates that the cardiac chambers exchanged sides and the entire heart is a mirror image of the normal situs. Conditions in which the anatomic type of the cardiac chambers cannot be determined are defined as *undetermined situs*.

Viscero-atrial Situs

Viscero-atrial situs refers to the interrelationship between the atrial situs and the visceral-abdominal situs. As pointed out by Van Praagh et al.,[31] the anatomic right atrium is almost always on the same side as the liver. In other words, there is side correlation between the atrial and visceral situs. But this is not a rule, as there are cases of atriovisceral heterotaxia[11] in which the right atrium is on the side of the body opposite the liver.

Inversion

Inversion implies that one or more cardiac chambers exchange sides. Hence, situs inversus is considered as inversion of the whole heart. Inversion may involve only one cardiac segment, as is seen in isolated ventricular inversion. In this condition, the anatomic right ventricle is on the left side of the septum, the anatomic left ventricle is on the right side, and the atria are in situs solitus. Other related conditions are isolated atrial inversion and isolated bulbar inversion (see Chapter 6).