SPECIAL ARTICLE

Pediatric Congenital Cardiac becomes a Postoperative Adult

The Changing Population of Congenital Heart Disease

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SUMMARY

The following commentary on the changing population of congenital heart disease will deal with: incidence and survival patterns—an overview; effects of diagnostic and surgical interventions on survival patterns—changing population; ultimate goals and aspirations—what are we trying to accomplish; types of surgical intervention—desirability of primary anatomic repair in infancy; and the problem of residua—setting the stage for future research.

Additional Indexing Words:
Congenital heart disease
Pulmonary hypertension
Ventricular hypertrophy
Ventriculotomy
Cardiac surgery
Myocardial hypoxia

The NATURAL history of congenital heart disease begins before birth, since most anomalies compatible with 6 months of intrauterine life permit live offspring at term. An impression of the overall problem can be derived from 1970 population figures. In that year, 3,718,000 live births were registered in the United States.1 An estimated 28,000 were born with congenital heart disease, i.e. 0.8%.2 About half died within the first year,3 leaving 14,000 survivors.

Those who succumb in early infancy generally have either complex anomalies or simple anomalies with severe circulatory derangement.4 It is now possible to perform palliative or corrective surgery on almost all congenital cardiac anomalies, even the most complex.5 Surgical management of the neonate and infant are undergoing radical changes, sometimes permitting one-stage repair rather than interval palliation. Survival patterns are affected, often profoundly. We are therefore confronted with a changing population of congenital cardiacs. These patients must be cared for, be it by a pediatric cardiologist who extends his or her interest to older subjects, or a medical cardiologist who has a satisfactory comprehension of congenital heart disease. Just as important are the primary physicians in the community who have a major responsibility for continued long-term care of patients with congenital cardiac disease, whether these physicians practice medicine or pediatrics. One purpose of this essay is to underscore the responsibility of training programs in meeting these needs which are already upon us, with every prospect of substantial increase.

Immense technical resources are at our disposal, permitting remarkably precise anatomic and physiologic diagnoses, and astonishing feats of palliative and corrective surgery. However, we are obliged to look beyond the present and define our ultimate goals. What do we seek to accomplish? The answer is clear. Our efforts should focus on the quality of long-term survival. The Congenital Heart Disease Study Group of the Intersociety Commission for Heart Disease Resources stated in its report, "The goals of long-term care are to bring the individual to the peak of his physical capability and to help him develop into a well-adjusted and effective member of society."6 In this context, let us examine the current and projected distributions in survival.
patterns of congenital heart disease. Some congenital cardiac valvular stenosis, which is functionally unimportant and not likely to progress with time. Others, such as mild valvular aortic stenosis, or functionally normal congenital bicuspid aortic valve, are of little or no physiologic significance in early life, but tend to become progressively stenotic as time passes. Furthermore, such valves are always at risk of infective endocarditis, which can convert a physiologically benign anomaly into a catastrophic mechanical fault. It is unknown whether the number of patients born with certain types of congenital cardiac valve disease will change. However, there is the prospect that increasing numbers of operated congenital cardiac will reach childbearing age and produce a higher incidence of offspring with congenital defects of the heart or circulation. Patients undergoing surgical intervention already represent an ever-increasing proportion of children who require long-term medical management because of surgical inoperability.4

When an inordinate amount of time, money, and personnel is expended to prolong the life of an infant for a few years, this effort speaks more for the level of technologic skill and dedication than for the real quality of the result. For such patients, early childhood is all there is of life. A few years of survival, imperfect in so many respects, may be a defensible objective in adults, but is a debatable goal in infants. Efforts yielding these short-term results should be looked upon as necessary milestones in developmental therapeutics, transitions on the way to more distant and desirable aims of normal or near-normal survival. Whether we are dealing with correction of congenital cardiac defects or developing rockets to the moon, we must not let the brilliant glare of technologic success obscure our long-term objective to promote the quality of life.

Before examining the problem of postoperative residua, we should define the general types of surgical interventions currently employed. Results depend upon the kind of congenital cardiac anomaly, as well as upon the type of surgical procedure (table 1). In establishing guidelines for the timing and type of surgical interventions, we might consider the following remarks as points of departure. Infants born without congenital cardiac defects are better off than those born with them.

Table 1

<table>
<thead>
<tr>
<th>Types of Surgical Intervention</th>
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<tr>
<td>1. Palliative surgery:</td>
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<tr>
<td>a. With anticipation of future correction, such as shunt operations for tetralogy of Fallot</td>
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<tr>
<td>b. Without anticipation of future correction, such as shunt operations for tricuspid atresia with pulmonic stenosis</td>
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<td>2. Anatomic correction:</td>
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<td>a. Without residua, such as division of a small patent ductus arteriosus</td>
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<tr>
<td>b. With residua, such as anatomic correction of tetralogy of Fallot</td>
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<tr>
<td>3. Physiologic correction:</td>
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<td>Such as Mustard or Rastelli operations for complete transposition of the great arteries</td>
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Given the presence of a significant cardiac anomaly at birth, the earlier it is anatomically corrected, the more nearly the patient resembles normal. It would seem, then, that an ideal goal is primary anatomic correction in infancy. Medicine is the only profession that strives to eliminate the cause of its own existence. Short of preventing congenital heart disease altogether, the best alternative is to recognize and deal with it as soon as possible after birth. However the siren voices in this apparent syllogism should not obscure the fact that no more than a handful of centers are currently ready to apply it.

The above remarks set the stage for an appraisal of the most important single problem that confronts patients after recovery from operation, namely the absence or presence of undesirable cardiocirculatory residua. The ideal of correction without residua is accomplished when an uncomplicated ostium secundum atrial septal defect is closed by direct suture, or when a nonpulmonary hypertensive patent ductus is divided, neither correction requiring ventriculotomy.

Let us now turn our attention to specific types of residual abnormalities present after anatomic and/or physiologic repair and look at selected examples in the context of the quality of long-term survival and the changing patterns of congenital heart disease. In this important area, more is unknown than known, so my main objective will be to pose questions rather than provide ready answers. Certain highlights will be touched upon; no attempt will be made to be comprehensive. In general, residual abnormalities following nonpalliative repair are those shown in table 2.
Table 2

Residual Abnormalities after Nonpalliative Repair

1. Residua after anatomic correction:
   a. Uncorrected defects (functionally normal bicuspid aortic valve with coarctation of the aorta or cleft but competent mitral valve with ostium primum atrial septal defect)
   b. Ventricleotomy
   c. Valvular regurgitation (valvotomy for aortic or pulmonic stenosis)
   d. Elevated pulmonary vascular resistance following repair of left-to-right shunts
   e. Prosthetic materials (patch for large ventricular septal defect, prosthetic valve replacement, homograft materials for anatomic repair of truncus arteriosus, etc)
2. Residua after physiologic repair — Mustard or Rastelli correction for complete transposition of the great arteries
3. Postoperative residual myocardial ischemia and ventricular hypertrophy

Residua after Anatomic Correction

Primary anatomic repair in the face of physiologically unimportant uncorrected defects is nicely represented by functionally normal bicuspid aortic valve with coarctation of the aorta and cleft but competent mitral valve with ostium primum atrial septal defect. There is little doubt that the natural history of unoperated coarctation makes the low surgical mortality a small price to pay. It is encouraging to learn from the National Heart and Lung Institute that age at operation—at least after 1 year—does not affect postoperative gradients across the zone of coarctation repair, and that early postoperative gradients, if present, do not progress when remeasured 10–15 years later. It is still the consensus that optimum age for correction of coarctation of the aorta awaits the seventh year, and that if vigorous medical management of congestive heart failure in infancy results in clinical stability, operation can be deferred to optimal age. On the other hand, primary repair in small infants is now selectively and successfully accomplished at a number of centers. Irrespective of the quality of correction, we do not know whether the risk of infective endocarditis (bicuspid aortic valve) or intracranial hemorrhage (berry aneurysm) will be affected. Upward of 25% of patients with otherwise uncomplicated coarctation have a bicuspid aortic valve7 which has been judged by Roberts to be perhaps the most common congenital anomaly of the heart or circulation8 (fig. 1). Although the natural history of a functionally normal bicuspid aortic valve is not fully known, Osler's emphasis on susceptibility to infective endocarditis still holds; furthermore, the bicuspid leaflets may thicken, calcify, and become obstructed as time goes on, or less commonly, evert, resulting in progressive aortic regurgitation.

Partial endocardial cushion defect is another case in point (fig. 2). The ostium primum atrial septal defect can be readily closed, but it is not yet known to what degree a cleft but competent mitral valve is susceptible to infective endocarditis or how well that valve, with its abnormal chordal arrangements, will remain competent in the presence of the degenerative diseases, changes in left ventricular geometry, or systemic hypertension of later life. The normal mitral apparatus is a complex, finely coordinated mechanism that requires for its competence the functional integrity of a number of anatomic elements working in delicate harmony.10 It is unlikely that the anatomically abnormal mitral valve of partial endocardial cushion defect will, in the long run, prove as durable as its normal counterpart.

Ventricleotomy is required for intracardiac repair of lesions such as large ventricular septal defects. Some complications of ventriculotomy are obvious and dramatic, such as postoperative ventricular aneurysm. On the other hand, alteration in the sequence of ventricular activation—bundle-branch block11 (fig. 3)—may have little functional significance. More to the point is the effect of ventriculotomy on the long-term performance of the incised ventricle. It is not known whether or how long such a chamber will perform normally or to what degree its functional reserve might be compromised when challenged by increased work or degenerative disease.

Direct operation without valve replacement in either valvular or discrete subvalvular aortic stenosis can virtually abolish the left ventricular-aortic gradient. Aortic regurgitation, together with valvular anatomic residua following valvotomy for aortic stenosis—assuming complete relief of obstruction—can be looked upon as a potential problem years after operation. In this context, three postoperative residua concern us, all of which may await adult life to express themselves. The first residual, mild aortic regurgitation following operation, may remain stable short of infective endocarditis or systemic hypertension later in life. Second, is the effect of relief of obstruction on predisposition to infective endocarditis; considering the high susceptibility of the unobstructed bicuspid aortic valve, it is unlikely...
that operation will have an ideal result in this regard, especially if the stenotic valve were intrinsically bicuspid to begin with, which is often the case. Nor should we forget that in discrete subvalvular aortic stenosis leaflet abnormalities and aortic regurgitation are common, so that the postoperative valve remains at risk of infective endocarditis. Third, a postoperative bicuspid aortic valve may have the same or theoretically greater tendency than the congenitally unobstructed bicuspid aortic valve to thicken, calcify, and become stenotic with the passage of time.

The problem of pulmonary hypertension, or more precisely, increased pulmonary vascular resistance in operative risk and postoperative results of left-to-right shunts continues to be an item of concern.12

**Figure 1**

Thoracic aortogram in a 10-year-old boy with coarctation of the aorta (arrow, linear zone of negative contrast distal to the left subclavian artery) and the typical angiographic picture of bicuspid aortic valve (large conjoined leaflet rather than three clear sinuses of Valsalva).
infants born with large ventricular septal defects, the high neonatal pulmonary vascular resistance exhibits one of four subsequent patterns: normal regression of the fetal pulmonary arteriole with a proportionate fall in resistance, delayed or incomplete regression, a rise in pulmonary vascular resistance after initial complete or partial regression, or rarely, persistence of the high pulmonary vascular resistance of the newborn. Relevant to this discussion is the patient with elevated pulmonary vascular resistance but a sufficiently large left-to-right shunt to warrant correction. Selection of such patients depends upon a question that is simple to pose but difficult to answer—namely, at what stage

*Figure 2*

Left ventricular angiocardiogram in a 6-year-old boy with a partial endocardial cushion defect. There is the characteristic angulation of the left ventricular outflow tract (gooseneck or swan's neck deformity). The mitral valve was only mildly incompetent. For comparison, a normal left ventricular outflow tract is shown from a patient with ostium secundum atrial defect. (Reprinted from Perloff, with permission.)
will removal of the stimulus of the left-to-right shunt still allow acceptable regression of pulmonary arteriolar disease? At what stage is the high-resistance arteriole still capable of anatomic involution? If the postoperative pulmonary vascular resistance does not adequately fall, pressure overload of the right ventricle persists and regression of hypertrophy is eclipsed. It is reassuring to learn from the Mayo Clinic that surgical closure of large ventricular septal defects in infants is not followed by late development of pulmonary vascular obstructive disease, which may be the case if the defect is closed later in life. The feasibility of primary intracardiac repair of ventricular septal defect in infants less than 1 year has been demonstrated by Sloan, Kirkin, Barratt-Boyes, and others.

Let us now turn our attention to the residua that may be associated with prosthetic materials. It is sometimes difficult to identify a specific effect of the prosthesis since insertion is likely to be accompanied by a number of other variables. For example, closure of a large ventricular septal defect requires a patch which is quickly incorporated into the ventricular septum and covered by a neointimal layer. Can we judge the effect of the patch alone in a patient in whom we must also consider the effects of right ventriculotomy, residual hypertrophy of the previously volume-loaded left ventricle and pressure-loaded right ventricle, and perhaps residual elevation of pulmonary vascular resistance? Jarmakani and co-workers have recently provided evidence that left ventricular contractile state is
depressed following repair of large ventricular septal defect after long-standing volume overload. Bloomfield implied that the presence of even a small defect in the ventricular septum might ultimately interfere with left ventricular performance. Since the septum is functionally part of the left ventricle in normal hearts and in ventricular septal defect, we can ask ourselves whether a patch incorporated into the septum interferes in the long run with the functional reserve of the left ventricle. I prefer to think not, but the answer is unknown, provided the question has been properly proposed.

Residua after Physiologic Repair

Residua after physiologic repair—as in complete transposition of the great arteries—poses problems that we have already touched on, such as effects of prosthetic materials; but it also poses problems of major theoretic interest, such as the long-term capability of an anatomic right ventricle to perform as a systemic chamber. Let us examine two types of repair, both of which represent great advances in promoting longevity in complete transposition of the great arteries—Rastelli and Mustard operations. Before specific comment, we should not lose sight of the fact that many infants with transposition now survive to reach physiologic corrective surgery because of prior creation of an atrial septal defect, clearly the most important palliative procedure in the treatment of such patients. The Rashkind balloon septostomy has been a major step forward in neonatal survival, even though some patients require later surgical septectomy to maintain improvement in systemic arterial oxygen saturation.

In 1969, Rastelli, Wallace, and Ongley described a method for complete physiologic repair of transposition of the great arteries with pulmonic stenosis and ventricular septal defect. The technic achieves redirection of ventricular outflows. The ventricular septal defect and aorta are connected by

![Figure 4](http://circ.ahajournals.org/)

**Figure 4**

Electrogram from a 5½-year-old boy who, 3 years before, underwent Mustard repair for complete transposition of the great arteries. An emergency balloon atrial septostomy had been done as a neonate. The electrocardiogram still exhibits right ventricular hypertrophy with a 20-mm R wave in V₁, indicating that the right ventricle continues to eject a systemic pressure.
a prosthetic tunnel so that the left ventricle becomes the systemic chamber; the pulmonary trunk is connected to the right ventricle by an aortic homograft and valve, so that the right ventricle becomes the venous chamber and the pulmonic stenosis is simultaneously relieved. Several concerns remain regarding long-term survival after this ingenious procedure; namely, the fate of aortic homograft and prosthetic tunnel, the effect on left ventricular function of using the ventricular septal defect as a permanent outlet, and the effect on right ventricular function of anastomosing the aortic homograft along the edges of the ventriculotomy.

The Mustard operation—in use about 5 years longer than the Rastelli—achieves redirection of venous inflows in transposition of the great arteries without pulmonic stenosis by inserting an intraatrial pericardial or Dacron partition. The partition directs pulmonary venous blood across the tricuspid valve into the anatomic right ventricle which continues to function as the systemic pump; systemic venous blood is directed across the mitral valve into the left ventricle which still functions as the venous ventricle. Physiologic correction is achieved, since pulmonary venous blood reaches the aorta and systemic venous blood reaches the pulmonary circulation. Ventriculotomy is not necessarily required since a ventricular septal defect, if present, can usually be approached through the tricuspid valve. Discounting late deaths caused by atrial arrhythmias and heart block, a postoperative problem of fundamental importance still confronts us in our quest for quality of long-term survival. The question simply posed is whether an anatomic right ventricle can perform as a systemic chamber as well as an anatomic left ventricle (fig. 4). The closest experiment of nature is congenitally corrected transposition of the great arteries with absent or functionally trivial coexisting anomalies (fig. 5). In both surgically corrected (Mustard) and congenitally corrected transposition, systemic venous blood flows across an anatomic mitral valve into an anatomic left ventricle and then into the pulmonary trunk, while pulmonary venous blood flows across an anatomic tricuspid valve into an anatomic right ventricle into an aorta. The coronary circulations to the systemic (anatomic right) ventricle are similar in both. Although the ideal natural experiment of uncomplicated congenitally corrected transposition is seldom met, some information is available that at least provides us with ranges if not with means. The X-ray in figure 6 is from an asymptomatic 37-year-old man with congenitally corrected transposition of the great arteries and mild left atrioventricular valve regurgitation. He was symptom-free, leading a normal life in all respects. Lieberson, Schumacher, Childress, and Genovese described a man who died at age 73 years with congenitally corrected transposition of the great arteries. Other examples have been reported of survival to age 54 and to age 60. On the other hand, Nagle, Cheitlin, and McCarty reported a 45-year-old woman who developed "spontaneous" failure of her systemic ventricle in the context of congenitally corrected transposition of the great arteries without systemic hypertension and with normal coronary arteriograms. Are these cases representative? Do they give us useful insights into the durability of the right ventricle as a systemic pump? The current consensus is still best expressed by a comment from Botem and Hultgren in their 1965 commentary: "Still unanswered . . . is the question of whether the right ventricle subjected to systemic work loads for many years would, for this reason alone, fail at an earlier age."

**Figure 5**

Schematic illustrations of congenitally corrected transposition of the great arteries and a normal heart for comparison. In corrected transposition, right atrial blood flows across a morphological mitral valve into a venous ventricle (VV) that is morphologically a left ventricle (LV) and then into the pulmonary trunk (PT). Left atrial blood flows across a morphologic tricuspid valve into a systemic arterial ventricle (AV) that is morphologically a right ventricle (RV), and then into the aorta (Ao). The left a-v valve is often malformed and incompetent (reversed arrows), and a ventricular septal defect is often present. (Modified after Schiebler et al., and published with permission, Perloff JK.)

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Residual Myocardial Ischemia and Ventricular Hypertrophy

Let me end this essay by dealing with two additional problems that await solution—myocardial ischemia in infants with congenital heart disease, and the fundamental issue of ventricular hypertrophy, its development and regression.

Ischemia concerns us from two points of view, namely, the unsolved relationship between ischemia and hypertrophy which will be touched upon below, and the effect of ischemic injury on ventricular performance after surgical repair. Tawes, Berry, Aberdeen, and Graham found a high incidence of myocardial ischemia in necropsy examinations of 103 hearts of infants who died after standard extracardiac operations for coarctation of the aorta, and tetralogy of Fallot (fig. 7). The ischemic changes were attributed to inadequate coronary perfusion of the grossly hypertrophied hearts of coarctation and to inadequate coronary perfusion coupled with hypoxia and polycythemia in tetralogy of Fallot. Similarly, Moller, Nahib, and Edwards called attention to scarring of papillary muscles in infants with severe aortic stenosis. The prevalence of ischemic injury to ventricular myocardium of infants with various forms of congenital heart disease is unknown, but its presence may influence both operative risk and long-term postoperative survival.
The development of muscle hypertrophy is a fundamental biologic principle. However, when adaptive ventricular hypertrophy fails to regress in the wake of anatomic and physiologic surgical repairs, myocardial reserve and life expectancy may be compromised. Hubert Meersen properly said, "Hypertrophy is undoubtedly the most important problem in the structural adaptation of heart muscle." I might paraphrase this statement and say, regression of ventricular hypertrophy is undoubtedly the most important problem in the structural return of heart muscle to normal following surgical correction. Much attention has been focused on the determinants of ventricular hypertrophy, but much remains unknown; less attention has been focused on the regression of ventricular hypertrophy, and a great deal remains unknown. Two basic questions can be posed. First, how does the myocardium convert pressure and volume information into biochemical information that results in fiber hypertrophy? Second, how does the myocardium translate relief from pressure and volume stimuli into biochemical information that results in regression of hypertrophy?

The myocardium utilizes energy produced by enzymatic degradation of substrates and stored in the phosphate bond of ATP. This chemical energy is converted into mechanical work. A relationship therefore exists between synthesis and utilization of chemical energy on the one hand, and mechanical work on the other. The structural adaptation to increased mechanical work is hypertrophy. We do not yet know whether the biochemical and ultrastructural equivalents of ventricular hypertrophy are the same in response to volume or pressure stimuli. Meersen pointed out that in rats swimming for as little as 2 hours, a slight adaptation swelling of mitochondria was seen with electron microscopy. Nair and his colleagues have shown that within 12 hours after experimental aortic constriction, there was an increase in RNA polymerase activity of nuclei isolated from the myocardium. Similarly, Page and co-workers found that after aortic constriction in rats, additional

**Figure 7**

*Interstitial fibrosis from the left ventricle of an infant who died after resection of coarctation of the aorta. (Reprinted from Ann Thorac Surg, with permission.)*
myofibrils, including polymerized actin in the thin filaments, were synthesized within 24 hours or less in response to the hypertrophic stimulus. In man, myocardial hypertrophy has been identified after 2–3 weeks of renal hypertension.

We must also define the role of myocardial hypoxia in the development of ventricular hypertrophy. For example, in anomalous origin of the left coronary artery from the pulmonary trunk, heart weights are, as a rule, greatly increased. Is hypertrophy in such patients (fig. 8) the result of a direct effect of hypoxia on viable myocardium, or the result of increased work of zones of myocardium remaining viable after infarction of other areas? Both mechanisms might be operative, but Hollenberg has shown that reduced $O_2$ tension results in increased rates of cell division and protein synthesis in cultured heart cells from the ventricles of chick embryos.

Once ventricular hypertrophy is established, the provoking stimuli can be removed, but little is known about regression of hypertrophy under these circumstances. Since conversion of chemical to mechanical energy is increased when pressure or volume demands are imposed upon the heart, it is reasonable to propose the converse, namely, that reduction in mechanical demands would be accompanied by an appropriate decrement in myocardial metabolism. Even if this were so, the critical issue is how such a change influences the structure and function of the hypertrophied heart, i.e., at what point in the development of hypertrophy can surgical intervention set the stage for regression that will allow the heart to function as a normal pump? It is noteworthy in this regard that the investigations of Skelton, Prindle, and Epstein showed that biventricular hypertrophy produced in cats by thyroxine administration was reversible, but

Figure 8
Electrocardiogram from a 15-year-old boy with anomalous origin of the left coronary artery from the pulmonary trunk. There are abnormal $Q$ waves in leads 1, aVL, and $V_5$. Posterobasal left ventricular hypertrophy is manifested by deep $S$ waves in leads 3, aVF, and $V_{6-5}$, with tall $R$ waves in $V_6$. (Reprinted from Perloff, with permission.)

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CONGENITAL HEART DISEASE

Despite regression of hypertrophy, papillary muscle function remained significantly depressed suggesting residual structural or biochemical derangement in those muscles. Adaptive hypertrophy is an expected feature of the fetal right ventricle which injects into the aorta at systemic pressure via the ductus arteriosus. After birth, with the stimulus of afterload removed, there is a gradual reduction in thickness of the right ventricular wall relative to septum and left ventricle. Does this change represent regression of right ventricular hypertrophy? Does each mammalian birth serve as a model to study regression of hypertrophy? Not so, according to de la Cruz and co-workers, who, in a quantitative study of the ventricles of normal children, concluded that both ventricles increased in thickness with age, but the increase was discordant because of a much greater rate in the left ventricle as compared to the right.\(^{45}\) Accordingly, the neonatal right ventricular wall does not undergo regression; it merely doesn’t increase its thickness as rapidly as the left. Should the fetal right ventricle continue to eject at systemic pressure after birth, as in cyanotic tetralogy of Fallot, then right ventricular wall growth keeps pace with left ventricle and septum, and long-term right ventricular performance is remarkably good.\(^ {46}\) Witness the relative infrequency of right ventricular failure in this anomaly.\(^ {18}\) On the other hand, if the right ventricle hypertrophies after normal adult ventricular wall ratios are achieved, the performance of that hypertrophied right ventricle is less adequate. Witness adults with mitral stenosis in whom the right ventricle fails at pressures far below systemic.

The electrocardiogram is often cited as evidence of regression of ventricular hypertrophy. Following relief of obstruction to right ventricular outflow, for example, a change from a tall monophasic R wave in V\(_1\) to an rSR’ coincides with a fall in right ventricular systolic pressure and is taken to represent regression of hypertrophy (fig. 9). More direct evidence of regression comes from determinations of right ventricular weights in hypoxia-induced pulmonary hypertension in rats. Abraham et al. have shown that when rats are exposed to chronic hypoxia in decompression chambers, they develop pulmonary hypertension and an increase in right ventricular weight.\(^ {14}\) Conversely, when the animals were removed from the hypoxic environment, the right ventricular weights returned toward normal. The same experiments showed that pulmonary vascular changes accompanying the pulmo-

![Figure 9](image)

*Figure 9*

Lead V\(_1\), pre- and postoperatively in a 6-year-old girl who had severe calcific pulmonic stenosis (gradient 95 mm Hg). The sequence illustrates electrocardiographic evidence of regression of right ventricular hypertrophy. A tall monophasic R wave becomes an rR’, rsR’ and finally an rSR’. (Reprinted from Perloff, with permission.)

nary hypertension regressed with removal of hypoxia.

In conclusion, the last two decades have witnessed the maturation of diagnostic and therapeutic skills in the management of congenital heart disease. The advances have been astonishing. Survival patterns have been affected, often profoundly so. We are therefore confronted with a changing population of congenital cardiaca; the pediatric congenital cardiac is becoming a postoperative adult. In this context, my essay has dealt with five related topics: natural incidence and survival patterns; effects of diagnostic and surgical interventions on survival patterns; ultimate goals and aspirations; types of surgical intervention; and finally, the problem of postoperative residua, especially the effects of hypoxia and the regression of ventricular hypertrophy.

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