Unusual Causes of Heart Failure

By Howard B. Burchell, M.D., Ph.D.

A DISCOURSE on unusual causes of heart failure naturally follows and overlaps any discussion on unusual causes of heart disease, and some repetition is unavoidable. However, one outstanding distinction is that in which the normal heart fails on exposure to a gross overload, as for example after the production of a large aortic-caval fistula such as has followed operation for an intervertebral disk.  

In any discussion on heart failure, some definition of the subject is mandatory. The term “heart failure” in this communication indicates a physical disability with a propensity for dyspnea, edema, and fatigability wherein the heart plays a dominant role, albeit perhaps not always as the central figure on the disease stage but, if not, as the director-producer in the wings.

In addition to the traditional two types of heart failure, namely the one in which the dominant feature is edema and overt venous congestion, and the other in which it is episodic pulmonary edema (frequently the two are combined), one must recognize two other clinical types. The first of these is exemplified by the ambulatory patient whose main symptom is marked fatigue, and the second, by the bed patient who presents the picture of pallor, apprehension, weakness, and perhaps a sense of breathlessness without change in respiratory effort. In the first category may be observed many patients having severe mitral insufficiency, and in the second, patients with advanced myocarditis; in the latter particularly, a hypotonic, hyponatreemic syndrome may be associated.

In the approach to unusual causes of heart failure, two worth-while classifications are given in tables 1 and 2. The traditional academic classification of disease into congenital and acquired varieties remains a basic approach, worth the effort of recall from student days (table 1). At another time, a parallel approach with focus sequentially on extrinsic and intrinsic causes may open doors leading to the recognition of previously occult causes of the failure (table 2). In many instances in which one may be puzzled by the nature of the heart failure, one may be rewarded by looking at a distance from the heart, because practically any organ system may be the chief malefactor and have both initial and continued responsibility in the various heart failure syndromes. In listing possible contributing causes (table 3), one cannot restrict with ease the conditions to rarities; rather, one can claim only that the causes are sufficiently unusual that they are not encountered regularly in day-to-day practice. It is only by constant vigilance that one will be alert to recognize the unusual and explore the possible etiologic ramifications. In recognizing the unusual, one may occasionally reap the reward of identifying a causal factor that permits of complete cure.

Congenital Causes of Failure

In the neonatal period, the child with heart failure may be suffering from one defect or a combination of many defects. Clues will be forthcoming from the auscultatory findings and the electrocardiogram, the size and shape of the heart, and the pulmonary vascularity in the presence or absence of cyanosis.

Out of the host of possibilities, one needs to mention specifically the rarity of total anomalous pulmonary venous connection, with obstruction to outflow either because of a single stenotic vein or a stenotic foramen ovale. Tricuspid atresia with a stenotic foramen ovale, premature closure of the foramen ovale, stenoses of the pulmonary veins, con-
Table 1

Traditional Academic Classification of Causes of Heart Failure

<table>
<thead>
<tr>
<th>Congenital causes</th>
<th>Acquired causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Neonatal, late postnatal</td>
<td>1. Traumatic (valve cusp)</td>
</tr>
<tr>
<td>2. Gross structural, myocardial</td>
<td>2. Chemical or toxic (emetine or endocrine therapy, alcohol?)</td>
</tr>
</tbody>
</table>

Acquired causes

1. Traumatic (valve cusp)
2. Chemical or toxic (emetine or endocrine therapy, alcohol?)
3. Inflammatory (infections, hypersensitivity states, granulomatous lesions)
4. Degenerative (nutritional, presbycardia)
5. Neoplastic (intracavity, myocardial, peri-cardial)

Table 2

Classification of Heart Failure from the Standpoint of Extrinsic and Intrinsic Causes

A. Extrinsic cause, heart basically normal
   1. Effect by dynamic overload
   2. Effect by myocardial injury

B. Intrinsic cause
   1. Gross mechanical (hydraulic) defect
      a. Valvular deficiencies, large shunts
   2. Gross myocardial deficiency
      a. Cause indeterminate, for example, familial idiopathic hypertrophy
      b. Cause determinate, for example, hemochromatosis, gargoylism

C. Combinations

Failure makes its appearance. Ebstein’s disease, though rare, should be remembered as a possible cause of heart failure in a young child. Here, the characteristic auscultatory, electrocardiographic, and roentgenologic findings should make the diagnosis readily apparent. In some instances, however, the right ventricle, being fairly competent, may result in a left-to-right shunt at the atrial level and produce a confusing picture.

A rare cause of heart failure, which may become clinically manifest in late childhood or early adulthood even though the defect is primarily congenital, is so-called corrected transposition of the great vessels, with insufficiency of the left atrioventricular (“mitral”) valve. One should know enough about the condition to be alerted to it when one encounters a young individual having mitral insufficiency, evidence of left ventricular hypertrophy, left atrial enlargement, and perhaps second- or third-degree heart block without a history of rheumatic fever. Another type of late failure related to a primary congenital defect is that occurring consequent to rupture of an aortic-sinus aneurysm, with its superficial simulation of a patent ductus arteriosus.

It may be emphasized here that occasionally subaortic stenosis occurs in association with pulmonary stenosis, with the ventricular septum being intact and the pulmonary stenosis dominating the picture and with the electrocardiogram suggesting primarily the pulmo-
to be prepared to correct it in addition to the more overt pulmonary obstruction.

In addition to the gross structural defects of congenital origin, there are myocardial functional aberrations such as defects of a biochemical nature, some of which are not as yet understood or identified. In gargoylism, an inborn error of metabolism produces deposits in the muscle cell, and heart failure occurs. In some families, sudden deaths have occurred in the siblings with few findings apart from slight enlargement of the heart. The routine methods of examination have not revealed the cause. In one such family under my observation, a girl showed a markedly prolonged Q-T interval in the electrocardiogram as the only evidence of abnormality, and the cause of this has remained completely obscure.

**Acquired Causes of Failure**

In this, the second category of heart disease, the usual subgroups of traumatic, chemical or toxic, inflammatory, degenerative, and neoplastic causes are briefly reviewed.

**Traumatic Causes**

Traumatic lesions of the heart with survival of the patient, but with heart failure, are rare. The most commonly observed are the penetrative lesions, with failure of the circulation related to acute tamponade. The possibility of such a problem occurring in an investigative laboratory from cardiac puncture deserves mention. My colleagues and I have seen one patient with traumatic rupture of the ventricular septum related to nonpenetrating trauma present with the picture of heart failure. Surgical repair having been accomplished, the heart has been returned to a normal functional state. I have not observed rupture of the aortic valve, but instances of rupture of the chordae tendineae of the mitral valve have been diagnosed, some of these occurring without known previous disease or identified trauma. A history of rather sudden appearance of dyspnea and a loud murmur, and the development of rapidly progressive heart failure, should orient one’s thoughts toward this diagnosis.
Chemical or Toxic Causes

A prime example of chemical, or toxic, lesions is that of emetine therapy, which, however, rarely causes gross heart failure. One might also consider in this subcategory excess endocrine therapy, as may occur with large doses of adrenal steroids or with overdoses of thyroid hormones.

Inflammatory Causes

In the subcategory of inflammatory causes are the subdivisions: (1) direct infections, (2) hypersensitivity state including possible autoimmune disease, and (3) granulomatous lesions. It is important to emphasize that the diagnosis of myocarditis, which, excluding rheumatic lesions, was frowned upon 2 decades ago, is now recognized as a not infrequent clinical entity, for example as a complication of viral diseases such as poliomyelitis, influenza, and the Coxsackie group of infections. These viral agents may be responsible for the initiation of heart failure "de novo" in some instances, but more frequently if valvular disease is already present. In the hypersensitivity group, certain patients with disseminated lupus present evidence of myocardial disease. Also in some cases of lupus, pulmonary vascular disease may develop and cor pulmonale may make its appearance. In the granulomatous group, in addition to types of idiopathic myocarditis, well-documented cases of sarcoid with gross myocardial involvement and heart failure have been reported. This syndrome has not been observed personally.

In recent years, there has been an apparent increased incidence of patients who have a relentlessly progressive type of chronic myocarditis, with the total duration of life after onset being some months or years. The clinical picture is often characterized by severe disturbances in rhythm and conduction, with heart block and paroxysms of ventricular tachycardia being characteristic. Late in the disease, the patient often has a persistent presystolic gallop rhythm and presents a shock-like picture already mentioned in the introduction as typical of some patients with heart failure and a low cardiac output. The clinical picture is essentially akin to that seen on occasion in years past, namely the picture wherein there has been a severe toxic myocarditis in diphtheria. Perhaps as an example, par excellence, of heart failure that is rare and is caused by myocarditis due to a specific agent is the failure related to Trypanosoma cruzi (Chagas' disease). Extensive personal experiences with myocarditis of varying types and causes have been recorded by Mattingly and Brigden.

A cause of heart failure that previously was uncommon but now is encountered with increased frequency is healed subacute endocarditis, the patient usually presenting with symptoms and signs of heart failure related to either gross aortic or mitral insufficiency. When both aortic and mitral insufficiency are present, it may be rewarding to consider the possibility that the aortic insufficiency was primary and the mitral incompetence secondary. This mitral incompetence may not be related to the usual location of destructive vegetations involving the contiguous portions of the mitral valve, or to rupture of the chordae tendineae, but rather due to a ruptured mycotic aneurysm of the anterior (or aortic) leaflet of the mitral valve. If one were fortunate enough to have perforations of both aortic and anterior mitral cusps, surgical cure of both the aortic and the mitral insufficiency is possible. Rupture of an aortic-sinus aneurysm simulating a patent ductus arteriosus has been mentioned, and at this time one may mention having seen 2 cases of syphilitic aortic aneurysm with rupture into the superior vena cava in one case and into the pulmonary artery in the other case; a continuous murmur and heart failure were present in both cases.

The syndrome of constrictive pericarditis in its characteristic form should be readily recognized, but when it is associated with a large cardiac shadow, a chronic effusion and a constricted epicardium, it is not easily differentiated from conditions of enlarged heart or other causes with chronic heart failure.
such as amyloid disease and endocardial sclerosis with a dilated left ventricle. Pericardial disease can persist for many years, producing the picture of chronic heart failure, and when it occurs in older people one may be lulled into a false sense of security by a previous diagnosis of hypertensive and coronary atherosclerotic heart disease. In some instances, one's suspicion of pericardial disease may properly lead to further investigation, utilizing such technics as cardiac catheterization, angiocardiography, or even direct surgical exploration. In regard to the preceding problem, tuberculosis as an etiologic possibility should never be forgotten, and this is true irrespective of the age of the patient. Rarer causes of chronic heart failure with pericarditis are chronic hemorrhagic effusions, slowly progressive metastatic malignancy, and cholesterol pericarditis.

"Degenerative" Causes of Failure

The term "degenerative heart disease" is justifiably in bad repute, but one can borrow it from the traditional subcategories of acquired disease and attempt to justify its use by defining the term as indicating a deleterious change in the function of the myocardium related to defects in its nutrition and in the functional integrity of its individual units. Among the nutritional inadequacies, one may place beriberi heart disease, which was reported on recently by Jones. Earlier studies indicated that a high-output type of failure was to be uniformly expected, but this has not proved true of all cases.

Occasionally, patients in the older age group with heart failure are encountered and one may be at one's wit's end to assign a logical etiologic diagnosis. While there has been a tendency, in large part related to statistical chance, to attribute the heart failure to hypertension in past years, or to coronary insufficiency, there may not be sufficient clinical evidence of such an etiologic background to be satisfying to an inquiring mind. To such a phenomenon, the term "presbycardia" has been applied, but one may consider this term only as a convenience, and it is difficult from my experience in cardioligic practice to define it and call it an established entity.

The possibility that some patients with heart failure have defects of the myocardium or the pericardium consequent to ionizing radiation cannot be ignored, but as a cause of failure such defects must be rare. In the general category of "degenerative" lesions may also be grouped the patients who are given the diagnosis of idiopathic hypertrophy, and these patients may or may not have some degree of endocardial sclerosis. One should exercise great caution in making the diagnosis of endocardial fibrosis in adult patients, even when these may present with a stereotyped syndrome including the features of chest pain of an anginal type, progressively severe heart failure, and systemic emboli. It is uncertain in such adult patients whether the endocardial fibrosis is or is not the primary lesion, and one may be properly dissatisfied with this term as a clearly defined etiologic diagnosis. Reports of endocardial sclerosis as it occurs endemically in the native African have engaged the attention of clinician and pathologist alike, but so far it is not clear whether or not the disease has an etiologic relationship to that seen in this country. Other types of apparently nutritional myopathies in the African also apparently occur.

To be mentioned also in the category of degenerative myopathies are those instances in which heart failure develops post partum, giving rise to the concept of "postpartum heart disease." I have not observed a pure instance of idiopathic postpartum heart failure, and one careful review has indicated that there are usually adequate reasons why the situation might have occurred, these reasons being related to specific types of heart disease or hypertension existing prior to childbirth. In this general area, however, must be mentioned the frequency with which idiopathic pulmonary hypertension has its onset in the immediate postpartum period. This naturally suggests that pulmonary emboli, that is, minute thrombi or amniotic fluid from the uterine cavity, has initiated the pul-

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monary arteriolar lesion. It is evident that another explanation must be forthcoming for other women not having such an onset and for men with this condition. The course of patients with idiopathic pulmonary hypertension usually runs from progressive effort disability to intractable right heart failure in a few years.

Neoplastic Causes

The traditional last subcategory in a classification of acquired disease is that of neoplastic lesions. Here one may think of primary and secondary neoplasms in the thorax and their location within the cardiac cavities, in the myocardium and in the pericardial space. The cases of atrial tumor have reached sufficient numbers that one cannot regard this as a particularly rare lesion, and one always hopes, if an individual presents with the picture of right inflow stasis or of mitral valve obstruction, to run into clues that may lead to the establishment of such a diagnosis and eventual surgical care.

In rare instances a tumor may invade the right atrium through the inferior vena cava and produce the picture of heart failure. Tumors of the kidney are the usual offenders in this regard. Primary tumors of the myocardium are rare, but may occur in infancy as rhabdomyosarcoma, with heart failure the presenting symptom. Secondary tumors of the myocardium, even though extensive in some instances, rarely lead to heart failure, and I have no example of such a case. Both primary and secondary tumors involving the pericardium are not really rare, and they produce not only the picture of acute tamponade, but also the picture of chronic heart failure of the congestive type. Of interest is the possibility that a mesothelioma occurring within the pericardium might present as a pericardial effusion, mild fever, and recurring arthralgias, simulating rheumatic fever on cursory introduction.

Contributing Causes of Heart Failure

Many of the contributing causes of heart failure mentioned in table 3 are not truly rare. Practically all organ systems are listed. Although conditions of the skin, reticuloendothelial system, fat depots, and gonads are not included, this does not signify that they have no importance for heart disease. Despite the fact that skin diseases are not responsible for heart failure, they may provide diagnostic clues such as café-au-lait spots, “spiders” and other hemangiomas, or more specifically, scleroderma or angiookeratoma corporis diffusum universale. In patients who have hypertensive crises, one usually thinks of pheochromocytoma; this condition presents a dramatic situation when a patient is admitted as an emergency to a hospital and the blood pressure is found to be in the neighborhood of 300 mm. of mercury systolic and 200 diastolic, and pulmonary edema is present. That such a state is sometimes related to transient ischemia of the brain may not be readily appreciated, but the presence of such a syndrome has been proved repeatedly in our clinic.

In hypertensive disease and heart failure associated with an arteriovenous fistula in the hilus of the kidney,13 there is an unusual wedding of 2 etiologic factors of failure, namely the hypertension and the high cardiac output. The mechanism of the high output as related to the fistula is self-evident, but the hypertension as it could be attributed to ischemia of the kidney distal to the fistula is of greater novelty.

In respect to Paget’s disease of bone, the evidence that there may be an arteriovenous type of connection in the involved bones is clear-cut, and in patients with extensive disease there may be a high cardiac output. My colleagues and I have observed the phenomenon of a high cardiac output as originally described by Edholm and associates,14 but as yet we have no instances of heart failure that can be primarily related to Paget’s disease.

Comment

In this review, a number of different approaches to classification of unusual causes of heart failure have been outlined, and a listing of unusual and perhaps not so unusual contributing causes by the various organ systems

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has been given. By definition, the unusual has been considered that which is of infrequent occurrence in day-to-day cardiologic practice, including that which often is unrecognized because of its uncommon and occult nature in the clinic but which, as later revealed at postmortem examination, often may not be so unusual. One has been tempted to reapply a previously discussed alphabetic or encyclopedic approach to the problem of heart failure, but it would seem better to identify the features of the failure and to follow ramifications of the specific clues to the various congenital or acquired conditions along the avenues of thought that are most promising. Herein, in regard to the congenital area, only a few interesting conditions have been listed, without any pretense to completeness or discussion of differential diagnosis.

Reference to unusual cardiac conditions may be had in the book published by Reich, the size of which also indicates the magnitude of the problem and the difficulty of differentiating the unusual from the usual. In the particular category of chronic myocarditis, additional emphasis is needed, as this condition appears to be becoming more common, the nature of the process is not understood and multiple etiologic agents may be operative. Along with Mattingly, I should like to stress the importance of cardiac arrhythmias and conduction defects in patients with chronic myocarditis. It may be mentioned that following the report that toxoplasmosis might be a cause of chronic myocarditis, the serum of our patients has been tested to indicate whether this infection might have been present, but the results have been negative.

**General Conclusions**

In the whole province of the discussion of the uncommon, there may be the aura of immodesty, pomposity, and sophistry. To be meaningful and have pragmatic values, a communication concerning the unusual should lead to better understanding of disease and to its control or cure. To have mentioned premature obliteration of the foramen ovale, with hydrops fetalis, which results in such early death, has no value for the practitioner, yet the condition has real significance in the attempt to understand a congested circulation and the mechanism of endocardial sclerosis. On the other hand, mention of cor triatriatum has practical value, as the thought of its possible presence might lead to the establishment of its presence and its surgical cure.

One is overwhelmed with the multitude of rarities that may be encountered, and there is a geographic distortion of what constitutes a rarity; for instance, endocardial sclerosis remains a rarity in this country, but is stated to be one of the most common causes of heart failure in Uganda among the native Africans. Likewise Chagas' disease has not been recognized as having been seen at the Mayo Clinic, yet I am told that in Brazil it is a common cause of heart failure; the medical student readily diagnoses it and suspects the disease on such slight evidence as right bundle block. In the approach to the unusual, one should avoid not only a geographic provincialism in diagnosis, in regard to colloquial labels, but also a "time provincialism," meaning the avoidance of parroting of diagnostic labels fashionable at the time.

There is no pretense to completeness in this personal appraisal of the problem, and only a few articles, a minute fraction of the available literature, which have especially piqued my interest are given as references.

The mass of data pertaining to rarities with which the medical scholar is confronted is so formidable that he may well fail in the solving of an assigned problem. The analogy, now fashionable, to a computer seems an apt one: the usual mind cannot integrate the findings, remember all the possibilities, make the necessary discriminatory matching of each sign and symptom with all the cases of each rarity in the world literature; in other words, both his biologic computer and the programming of it seem inadequate. Indubitably, there will be a place for electronic computers in the diagnosis of the unusual in the future.

A medical rarity may serve as a true test
of a physician, as measured by his answer to a question: when he is confronted with a difficult problem in his specialty and has overlooked or disregarded signs that suggest a rare condition susceptible of specific therapy and cure and that have been thought to be diagnostic by another consultant, can he affirm, "I hope that my colleague is right and that the patient's life will be saved, rather than that he is wrong and that my reputation will be saved"? (A tinge of hope for saving of the reputation is admittedly justifiable.)

**Summario in Interlingua**

Es presentate un revista de causas inusual de disfattimento cardiae. Le termino "disfattimento cardiae" es definito como designante un stato de invaliditate physic con un propensiitate pro dyspnea, edema, e fatigabilitate in qu le corde ha un rolo, si non central, al minuo dominante.

Es discutite le valor complementari de duo classificaciones del causas inusual de disfattimento cardiae. Le un distingue causas congenite ab causas acquirite, durante qu le altere estabil gruppos distintec de causas intrinsec e extrinsec. Le duo classificaciones es presentate in Tabulas 1 e 2, supplementate in Tabula 3 per un lista de causas contributori.

Le autore organisa su detaliate discussion del causas inusual de disfattimento cardiae in le gruppos de causas congenite e acquirite, con un subdivision del secunde in causas traumatic, chimie e toxic, inflammatore, degenerative, e neoplastics.

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