

# Cardiac Surgery for Congenital Heart Disease in a Multiproblem Family

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**Dr. Burr** (*Associate Professor of Family Practice*): Today's presentation and discussion of a case of major cardiac illness in an individual patient in a multiproblem family will initiate a new approach to a clinical discussion which integrates psychosocial factors with disease processes on a family basis. This departure from the traditional grand rounds formats supports the basic philosophy of the Family Practice movement. Rather than viewing the patient in terms of an organ-centered illness, the Family Practice Grand Rounds will examine the pathophysiology of the illness in relationship to those intra- and inter-personal factors which have a

major impact on our therapeutic efforts. The family dynamics frequently are keyed to the successful intervention of an illness, and most certainly the family homeostasis is disturbed during and after a major illness. To give us a better understanding of the family setting prior to the major illness, Miss Theresa Arciniega, a member of the Family Practice team, will describe the family setting.

**Miss Arciniega** (*Medical Social Worker*): The family consists of four members: father and mother, age 25; daughter, age 4; and son, age 1½ years.

The father comes from a family of six. His father is 68, his mother is 49 years of age, and they are presently separated. His three brothers are ages 30, 27, and 21, and each has achieved considerable social and economic success. He comes from a competitive family. Being unemployed, he is considered the "Black Sheep" in the family. He responds to this judgment by saying he is glad that he is poor and not as materialistic as his father, mother and brothers are. He has completed one year of college credit, receiving at that time straight A's.

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The father has worked as a railroad surveyor, gas station attendant and has held other laboring jobs. As a railroad surveyor, he grossed \$1400. per month, but terminated that job under pressure from his wife, who resented his long absences from home. His job history shows frequent difficulties with authority figures. He has been unemployed for two years, and the family is currently receiving Aid to Families with Dependent Children (AFDC).

The mother also comes from a family of six. Her father is 53, mother 41, brother 25 and sisters 24 and 21 years of age. She comes from a close family typical of her Mexican-American cultural background. After completing high school, she entered Beauty School, but dropped out to get married before finishing her training.

The couple has been married for five years. It is the first marriage for each. They appear to have relied heavily on each other for support, and have made few friends. The marriage relationship has been frequently strained, however, and neither partner has shown effective verbal communication when problems arise. The mother has been generally depressed for three years, with frequent crying spells, lack of motivation and malaise. She has been frustrated by not understanding why she has been often unable to perform her usual household tasks. The father has presented with a variety of psychosomatic complaints over the past several years, including hyperventilation episodes and "blackout spells" without demonstrable underlying cause.

The family has utilized multiple specialty clinics, drop-in clinic and the Emergency Room at Sacramento Medical Center. More recently they have received care in the Model Family Practice Clinic, but often fail to keep scheduled appointments.

**Dr. Burr:** Having summarized the family's present status and background, we will now turn to the major clinical problem of the mother, which will be presented by her family physician, Dr. William Dabney.

**Dr. Dabney (Family Practice resident):** This 25-year-old white married woman was essentially well until approximately 3 months ago when she began developing chest pain. The pain was intermittent until about 3 weeks prior to admission. The past three weeks she had almost constant chest pain. This chest pain has been in her anterior chest lasting only for a few minutes with radiation down both arms, especially her left arm. Exertion seemed to increase the pain and on the morning prior to admission this occurred as she got out of her bed. The patient denies nausea, diaphoresis and only complains of a slight shortness of breath. She also has developed orthopnea and dyspnea. She denies having any chest X-rays prior to November 1972. At that time she was noted to have cardiomegaly on a routine chest X-ray for a job physical. She denies any other cardiorespiratory symptoms especially hemoptysis, pneumonia, asthma, prior history of paroxysmal nocturnal dyspnea, heart disease, shortness of breath, congenital heart disease or knowledge of heart murmur.

The patient's review of systems is entirely non-contributory except for the genito-urinary tract. She admits nocturia recently with considerable frequency but denies urgency or burning. Patient is a gravida 2 para 2 AB-0 without problems concerning her menstrual periods, but she does admit to stress incontinence which apparently started before her pregnancies. She was hospitalized for "blood clots" in her legs while on birth control pills 2 years ago. She was hospitalized in August 1972 at Sacramento Medical Center for ocular muscle surgery. She takes no current medications, denies other serious medical problems and has a non-contributory family history.

Initial physical examination showed an obese white female in no acute distress. She was afebrile, with blood pressure of 100/76, pulse 76 (regular) and respirations 22. The only positive physical findings were a grade 2/6 systolic in-

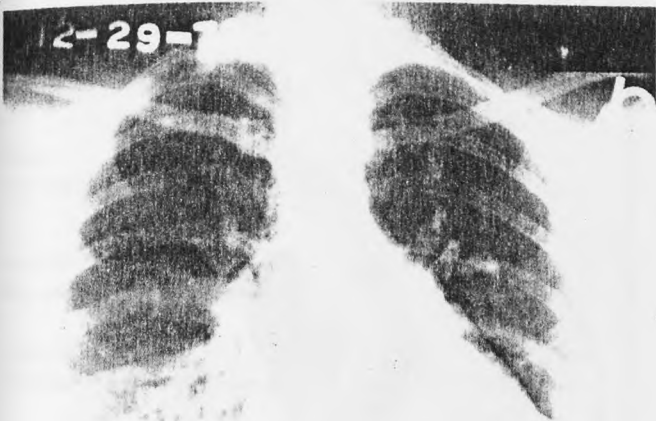


Figure 1

jection murmur at left sternal border and S 2 with fixed split with a loud P2. The cardiac PMI could not be seen or palpated due to obesity. There was no organomegaly, neck vein distension or peripheral edema. Lungs were clear to auscultation and percussion.

Initial laboratory data included the following:

- WBC 9200 (with normal differential)
- HB 14.6 gm% : HCT 42.3
- UA Within normal limits

Chest X-ray: Showed a prominent pulmonary artery and right ventricular outflow tract, probable right ventricular enlargement and possible left ventricular enlargement; lung fields were clear (figure 1).

EKG: Showed incomplete right bundle branch block (figure 2).

The tentative problem list was as follows:

1. Chest pain, etiology to be determined; rule out pulmonary embolis, rule out pulmonary hypertension.
2. Cardiomegaly
3. Obesity
4. Status postoperative for left lateral and medical rectus resection for left exotropia with a left hypertropia.

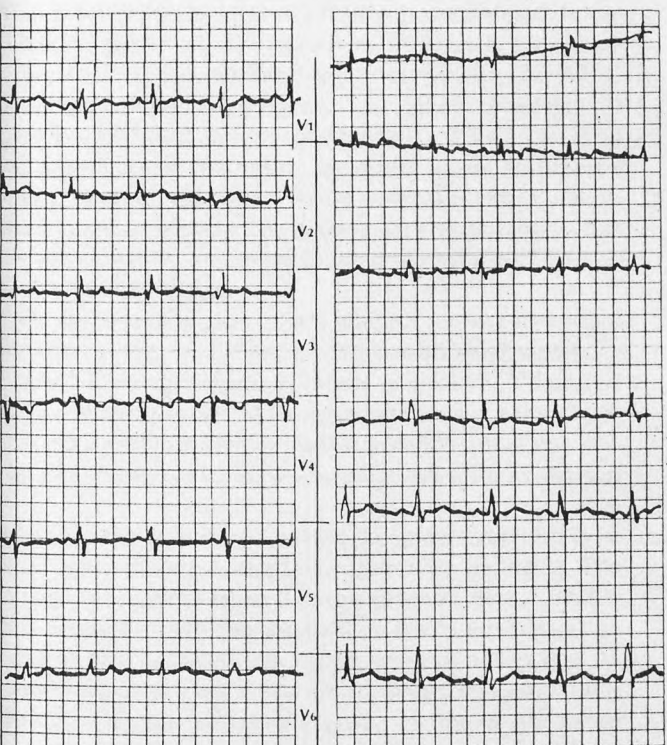
The initial plan was as follows:

1. Arterial blood gases, 6/60 and 12/60.
2. Heparinization with intermittent heparin therapy monitored by ACT.
3. Lung scan as soon as possible.
4. Cardiology consult STAT.

Arterial blood gases were reported as: PO2 85, pH 7.46, PCO2 31 and HCO3 21.5. Lung scan was normal. The patient was started on Heparin (7,000 units every 4 hours) and Coumadin 10 mg per day. Cardiology consultation concurred with the possibility of pulmonary embolism, but suggested right heart catheterization. On December 31, 1972, two days after admission, the patient had a severe episode of chest pain during the night which was unassociated with EKG or chest X-ray changes. The patient was shifted to maintenance doses of Heparin and Coumadin. A venogram of the right leg was performed and reported as normal. Catheterization of the right and left heart was performed on January 8, 1973. A diagnosis of atrial septal defect was made by passing the catheter from the right atrium into the left atrium at a high level. The O2 step-up in the right atrium was diagnostic and showed a shunt of probably 2.5:1, a pulmonary artery pressure of 35-40, but no other defects were found. On the morning after the heart cath the patient began to have an episode of severe chest pain, crushing in nature, that seemed to take her breath away but with marked relief with nitroglycerine. She had two subsequent episodes that day, the last being 2 p.m. but none following that. The patient did very well through the night and on the 10th of January it was elected to discharge her, but first, to digitalize her. This was done and it was elected to readmit her on the 24th of January for surgery. She was discharged with a diagnosis of:

1. Congenital heart disease — atrial septal defect
2. Congestive heart failure — early and mild
3. Chest pain secondary to No. 1
4. Probable angina pectoris, mild

Figure 2



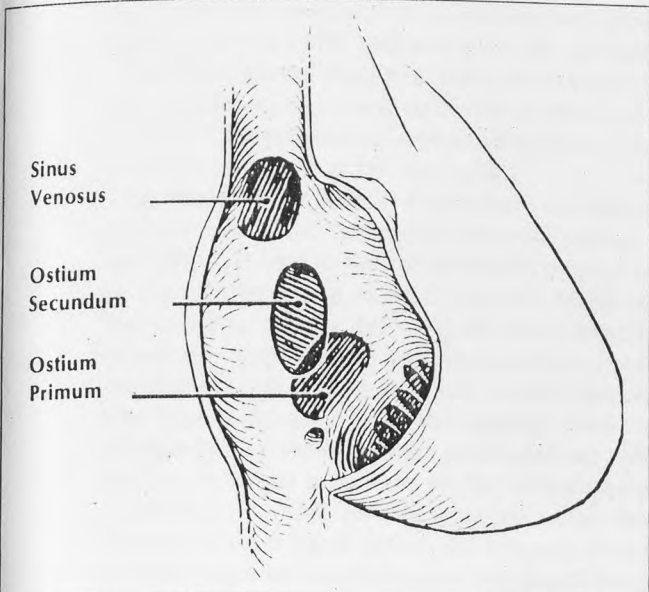


Figure 3

Patient was discharged home on digoxin, 0.25 mgs. per day. The patient was seen on the 17th of January in the Family Practice Clinic for follow up and it was noted at that time that her congestive heart failure seemed to be stabilized with markedly decreased dyspnea on exertion and no evidence of paroxysmal nocturnal dyspnea. There was no pedal edema at that time. Chest was clear; the heart rhythm was regular and the murmur was unchanged. It was decided the patient should return on 24 January for admission for work-up for cardiac surgery on January 26, 1973.

**Dr. Iben (Professor of Surgery):** The patient was taken to surgery as scheduled. Under total bypass, a 2.5 x 4 cm. atrial septal defect of the secundum type was found and closed primarily without difficulty. There were no other abnormalities found at surgery. There were no complications during or after surgery. She was discharged doing well on the sixth postoperative day.

There are several major points which I feel are important to be made in this case. First, a patient with atrial septal defect and a left to right shunt in excess of 1.5:1 should preferably have surgical repair of this problem during early childhood before entering school. In this instance, there was failure by any physicians to make the diagnosis until 25 years of age.

Secondly, this patient illustrates the most common type of atrial septal defect — the secundum type (figure 3). The least common variety treated is the primum defect. This is usually located above the portion of the heart formed by the endocardial cushions. An ostium primum defect may be associated with the cleft mitral and/or tricuspid valve. If it is associated with the mitral, the aortic leaflet of the mitral valve is involved by a large cleft and if it involves the tricuspid valve it is usually in a narrow leaflet. Occasionally, there is a ventricular septal defect present as well as a primum defect and if these two septal defects are present with cleft

mitral and tricuspid valves, one encounters an AV communus defect. The common associated anomalies are pulmonary stenosis in which case the patient may appear cyanotic and have what is described as a tetralogy of Fallot. Another associated anomaly is persistent left superior vena cava.

Thirdly, the history with atrial septal defects can be quite vague. There may be a history of minimal heart murmur that some physician has described during the patient's earlier years, or it may run the gamut of symptomatology running all the way to episodes of frank cardiac failure or atrial arrhythmias.

Fourth, there is a common pattern of typical findings in patients with atrial septal defects. The physical signs which are encountered are evidence on examination of right ventricular hyperpathy. The murmurs are of relative tricuspid and pulmonic stenosis. Unless the patient has a concomitant pulmonary valve stenosis, or has developed intense pulmonary vascular resistance, the patient will not be cyanotic. The X-ray appearance is quite routinely one of right atrial and right ventricular hyperpathy and evidence of main pulmonary artery predominance. Electrocardiographically, there is evidence of right ventricular hypertrophy with complete or incomplete bundle branch block. Patients who have sinus venosis defects may show coronary sinus instead of sinoatrial pacing. This is evidenced by inverted P-waves in leads 2, 3 and AVF and evidence of improper formation of the sinoatrial area during embryology. It is important to recall that although secundum defects may show left axis deviation on occasion, patients with primum septal defects always show this particular change on the electrocardiogram. From age 25 on, the patient is susceptible to paroxysmal atrial tachycardia, atrial flutter, and atrial fibrillation. These particular defects are apparent in this order as they get older.

Fifth, it is important to point out that cardiac catheterization must be performed on all of these patients preoperatively so that the type of defect can be more carefully mapped out and delineated for the surgeon. However, at times it is very difficult to know precisely the type of defect or the associated anomaly until the heart is observed during the operation. For this reason during the exposure of the heart, we stepwise look for anomalous veins, a persistent left superior venacava and, of course, for the thrill of mitral or tricuspid insufficiency that one might see in primum defects. Almost all secundum defects can be closed primarily without the addition of a teflon or pericardial patch. Sinus venosis defects without anomalous pulmonary return may be closed primarily but with the anomalous pulmonary return these require a pericardial patch which places the veins and the septal defect on the left side of the septum. In the repair of a primum atrial septal defect a woven teflon patch is usually utilized for its closure. This is of course done only after the cleft of a mitral valve is repaired.

And finally, it should be noted that the mortality associated with surgical management of atrial septal defect is ex-

tremely low. We can expect one surgical death in 5,000 patients with the sinus venosus or secundum variety and one death in 2,000-2,500 patients with the primum defect.

**Dr. Burr:** Having dealt with the surgical aspects of our patient's problem, it is now pertinent to focus on rehabilitation. Dr. Joseph Bonanno will describe how this part of the patient's care should be managed.

**Dr. Bonanno** (*Assistant Professor of Medicine*): Optimal medical care is the basic foundation of cardiac rehabilitation. The first step in the rehabilitation process then would be the assurance that optimal care was being rendered. In this case, the surgery itself was the most important part of her cardiac rehabilitation. In some instances, cardiac surgery can restore patients to a relatively normal state so that once they have recovered from the surgery, they can then be handled in the same manner as any patient without heart disease. In other instances, severe limitations persist and continuous medical management becomes necessary. In the case of atrial septal defect, the amount of medical management required depends upon the age of the patient and the extent of complications present prior to the surgery. This particular patient had a relatively small left to right shunt and normal pulmonary artery pressure and hemodynamically speaking, she should experience a complete cure from her surgery. She will have a small liability to develop bacterial endocarditis, atrial fibrillation and systemic embolization.

In addition to good medical care, the second most important step in the rehabilitation process involves informing the patient. Incredible as it seems, there are a large number of patients who have undergone open heart surgery without ever knowing what the nature of their problem was, how it was repaired or what to expect in the future. If you expect the patient to have residual impairment he should be cautioned as to his limitations. If on the other hand you expect complete recovery, both the patient and the family need to be reassured that that is the case. It is a common experience to find patients with a cardiac neurosis because someone in the past had told them that they had a "cardiac murmur" without ever adequately explaining what this meant. Just think of the potential for developing a cardiac neurosis that would be present in a patient such as this woman for her to retain an image that she had a "hole in her heart" which may not have been fully corrected — she should be told that she is cured.

The next step in the rehabilitation of this patient involves the activity prescription during the convalescent period. Since her pre-existing problem was mild and a total cure can be expected, her convalescent management should not differ from any other patient with a thoracotomy. If she had had more extensive disease pre-operatively, her resumption of activity during the convalescent period would have to be retarded. The patient's ability to return to work subsequently depends upon the severity of the disease pre-operatively, the effectiveness of the surgery, the number and type of surgical complications, if any, and the type of work to be re-

sumed. When their exercise tolerance or ability to work is in question, their work capacity can be tested with a progressive, multistage stress test using a treadmill or bicycle ergometer. If stress testing is required, it should not be performed before the 6th post-operative week. Physical deconditioning would be expected to occur as a result of the inactivity associated with the post-operative state and consequently the cardiovascular functional capacity would be underestimated if this test is done prematurely. Patients with residual impairment can almost always improve their exercise capacity through the employment of a carefully guided exercise training program. Such a program, however, must be very carefully spelled out with the exercise prescription derived from the results of the progressive multistage stress test.

**Dr. Burr:** With the patient's rehabilitative plan now underway, it is useful to again focus on other aspects of the continuing care of the whole patient as well as the impact on the family of this major illness.

**Dr. Dabney:** The patient was discharged from the hospital after repair of her atrial septal defect on February 2, 1973. She was seen twelve days later in the Model Family Practice Clinic for her first follow-up visit. She was feeling much better and had no recurrence of chest pain or dyspnea. The patient complained only of *headaches* (occipito-frontal) which were felt to be *tension* headaches. The patient stated that there had been a lot of tension at home with her husband out of work. She also complained of *heartburn* which was rapidly relieved by Tums. This was felt to be a mild peptic esophagitis and treated with Mylanta. Examination was completely within normal limits except for some scattered rales that cleared with coughing. She was given a 1,000 calorie diet.

The patient has continued to do well on subsequent post-operative visits. She is now three months post-surgery, has no chest pain, dyspnea or pedal edema, and has an improved exercise tolerance. She has resumed all normal activities including hiking in the mountains.

The family has been seen at home on several occasions after surgery by Miss Arceniga, who can further describe what changes have been recognized in the family.

**Miss Arceniga:** Several major patterns can be recognized in the family unit during the post-operative period. The patient's husband presented several times in the Emergency Room shortly before and after her surgery, with multiple complaints for which no organic basis was found. On one occasion, he complained of nervousness, dizziness, weakness and nausea. On another occasion he had vaguely described chest pains. On still another occasion, he presented a picture of acute anxiety reaction with a complaint of "feeling messed up in my mind."

The post-operative period was featured to keep appointments in the Model Family Practice Clinic and frequent drop-in visits to the Emergency Room, including visits for the children (for such problems as otitis media and constipation).

Meanwhile, the father continued to show lack of motivation to gain employment or assume a leadership role within the family. He was unable to make contact with his Social

Service worker after several half-hearted attempts. The Work Incentive Program was described to him, but he failed to explore this alternative. He talked of moving to an isolated rural area with some friends, but gave up these plans due to his wife's disapproval. A tense and uncommunicative relationship has developed between the couple — she seems to doubt his motivation for work. It appears that she is more concerned than he with their future plans and needs, and is considering a possible clerical job.

### Discussion

**Dr. Joe Tupin** (*Professor of Psychiatry*): As we review the family's visits to the hospital and the emergency room over the last several years, a number of things suggest that the family is experiencing psychosocial difficulties. The mother has presented a number of times with such complaints as heartburn, headache and fatigue. These complaints are often stress related, and have occurred after her other surgeries.

In the father's history, we notice admissions in 1969 for a dislocated arm and again in 1972. This is followed by a fracture of the great toe in August of 1972, and I begin to wonder whether he is accident prone, perhaps as a function of stress. Shortly after his visit for his fractured toe, he was seen for a "blackout spell" and then within two days he was again seen in the Model Family Practice Unit with a complaint that his "mind is messed up." While his wife is admitted to the hospital for her cardiac evaluation, he is seen in the emergency room with complaints of chest pain. He was then seen by a neurologist and psychiatrist and thought to have a psychosomatic problem. This would be quite in keeping with the fact that his wife, at that point, is being evaluated for chest pain, and would strongly suggest his anxiety which leads to such a complaint is related to her hospitalization and he, in fact, is identifying with her complaint. This further suggests an unusually dependent relationship between the two.

In looking at the children's complaints, we early find a fair number of upper respiratory infections, which seem quite consistent with their age. However, beginning in August of 1972, the daughter complains of constipation, an uncommon complaint for children. After their mother's operation, there are several visits in March of 1973 where more ill-defined upper respiratory complaints such as "ear pain" and "recurrent cough" occur. This may reflect some beginning pattern of somatic expression of tension or more likely it reflects the limited capacity of the parents to deal with minor problems without help. It is also known that these children have failed to thrive and that raises the serious problem of the adequacy of the parents' care for them. It would appear that the family has not functioned as an effective unit for a long time.

The concept of the "multiproblem family" has been widely noted. This term appears to characterize this family in terms of its clear-cut medical, psychiatric and social problems. This family appears to be disorganized, failure-oriented and in a downward spiral. Firm social intervention is needed by appropriate community agencies. I doubt that family counseling or therapy alone would be effective at

this point. Families such as this cannot be dealt with on a piecemeal basis, but rather must be addressed as social, psychological and medical problems on a family basis. This, it seems to me, is the "problem case" of Family Practice.

**Dr. John Geyman** (*Professor of Family Practice*): There are several important points to be made in further discussion of this Family Practice Grand Rounds. This presentation has clearly shown that, in addition to the individual patient, the family as a dynamic unit is also our patient. This perspective is central to Family Practice. In this instance, for example, it allows the family physician and his team to better recognize, understand and manage recurrent psychosomatic and behavioral problems in husband and wife. It also has predictive value in terms of other potential future medical problems within the family, such as severe marital conflict in the couple, enuresis or school phobia in the children. The present state of family disorganization can be expected to play a major role in the development of all future health problems for all family members.

Major critical events, such as birth, major illness or disability, can be expected to substantially alter previous family dynamics and cause some reorganization of the family unit — such reorganization can be in either a positive or negative direction. In this instance, cardiac surgery as a critical event appears to have contributed to the continued deterioration of the family unit, and has unfortunately not resulted in constructive role changes in the patient's husband. The family physician can learn much about the family by monitoring its response to critical events.

This multiproblem family severely challenges all health professionals involved in its care. Optimal management of specific problems will frequently not be fully effective despite the skilled efforts of the Family Practice team, consultants and other community resources. The family physician should recognize these difficulties and not become unduly frustrated for he remains the family's entry and major access to health care to the extent that it can be effective.

The type of presentation illustrated by this Family Practice Grand Rounds appears to have a number of useful advantages as a teaching technique by illustrating the concepts of Family Practice in a clinical context. Such principles as the following can be demonstrated:

1. Integration of behavioral science with organic disease.
2. Continuity of comprehensive care of the whole patient involves all stages of care — preventive and health maintenance, early diagnosis of asymptomatic disease, care of symptomatic disease, rehabilitation and care of terminal illness.
3. The family unit as a dynamic and evolving milieu for health and illness of all its members.
4. The necessity of a team approach to the care of families.

Family Practice Grand Rounds facilitates a multidisciplinary approach to diverse kinds of problems seen by the family physician and offers a useful approach to learn more about the family in health and illness.

