CLINICAL PROBLEMS IN CARDIOPULMONARY DISEASE

Idiopathic Hypertrophic Subaortic Stenosis

Clinical Evaluations by Eugene Braunwald, M.D. and E. Douglas Wigle, M.D.

CASE SUMMARY
James C. Dillon, M.D.*

The patient is a 36-year-old white woman, who has been observed in our cardiology clinic since 1968, with the diagnosis of idiopathic hypertrophic subaortic stenosis (IHSS). Her symptoms were vague, consisting of fatigue and shortness of breath in 1968. There was no chest pain at that time. Review of this patient's family history failed to reveal any other member with IHSS. The chest x-ray film at that time was interpreted as within normal limits. An electrocardiogram was also normal. Echocardiography revealed no outflow obstruction. In 1968 the patient underwent cardiac catheterization, at which time she was found to have normal right atrial and pulmonary artery pressures, with a slightly elevated pulmonary capillary wedge pressure of 14 mm Hg. Left ventricular pressure at that time was 140/16 mm Hg, with no resting pressure gradient across the left ventricular outflow tract. With exercise, a gradient of 5 mm Hg appeared, and with isoproterenol administration, the gradient increased to 110 mm Hg. Cardiac output was 5.5 liter/min, with an index of 3.2 liter/min/M².

After the initial evaluation, the patient was given a trial of propranolol therapy. This drug was continued for three months, with amounts increasing to 160 mg daily in divided doses. She felt that the propranolol made her symptoms worse, manifested by an increase in the incidence of shortness of breath as well as insomnia. For this reason, treatment with propranolol was discontinued and she has had none since that time. Recently, there has been a marked change in the patient's symptomatology, with increasing shortness of breath and fatigue and, in addition, for the past 12 months, she has developed chest pain. The pain has been described as "an exertional substernal pressure which radiates to her jaw." It has at times occurred after eating. In 1972, her chest x-ray film disclosed a marked cardiac enlargement, compared to previous examinations. Her electrocardiogram is also dramatically changed, showing an abnormal left axis as well as marked QRS voltage and ST-T wave changes. Echocardiography now shows a marked degree of left ventricular outflow obstruction at rest, with hypertrophy of the interventricular septum.

Her present physical examination has not basically changed since 1968. Her blood pressure is 130/70 mm Hg, with a pulse rate of 80/min and regular. Cardiac examination reveals the apical impulse to be in the fifth intercostal space in the mid-clavicular line. There is a sustained left ventricular lift. There is a grade 3/6 systolic ejection murmur heard at the left sternal border, radiating to the base and the apex, and increasing markedly with Valsalva's maneuver. The remainder of the physical examination was normal.

On Nov. 15, 1972, the patient underwent cardiac catheterization. At that time she was found to have normal right atrial pressures and a pulmonary artery pressure of 32/13 mm Hg. Pulmonary capillary wedge pressure was 20 mm Hg. Left ventricular pressure was 170/24 at the apex, and 112/26 mm Hg high in the left ventricle, resulting in a resting pressure gradient of 58 mm Hg. With exercise, the gradient increased to 84 mm Hg, and the patient was not given isoproterenol. Selective cineangiograms were compatible with hypertrophic subaortic stenosis.

In summary, we have a 36-year-old woman without familial history of IHSS who, over a period of four years, has shown marked progression in her illness.

QUESTIONS TO BE ANSWERED
1) What should be done at present?
2) Should she be given propranolol again and observed by objective testing to see if her outflow tract obstruction decreases?
3) Should this patient be subjected to operation?
4) If an operation were recommended, what type would you suggest?

Comments by Eugene Braunwald, M.D.*

Since the patient did not have obstruction to left ventricular outflow at the time she was first given propranolol, it is not surprising that she derived no benefit from this drug. The situation is different now; there is moderately severe obstruction at rest,

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and the outflow pressure gradient rises further with exercise. Therefore, it would be my recommendation that she be given another trial of propranolol under careful observation. In view of the difficulty that she experienced earlier, I would recommend starting with a small dose, i.e., 10 mg, qid, with frequent clinical observation and exercise testing as described elsewhere\(^1\) with gradual escalation of the dose.

If she does not derive major symptomatic improvements from propranolol therapy, as only a minority of patients do, then I think that operative intervention is indicated. A variety of operative approaches have been proposed, and many of these have been discussed in a recent international symposium on IHSS.\(^5\) In my experience, the operation devised by A. G. Morrow which consists of a major myotomy and minor myectomy of the outflow tract has much to commend it.

On the one hand, it is relatively conservative in that the incision into the ventricle is made through the aorta rather than through the left ventricle, the right ventricle or left atrium (other more radical approaches) and in that relatively little muscle is removed. However, this procedure has been shown to be remarkably effective, and the risk is relatively low,\(^4,3\) less than 10 percent in severely symptomatic patients. The deep incision in the outflow tract, coupled with the removal of small amounts of myocardium has resulted in the total or virtual abolition of the obstructive gradient consistently, and what is even more encouraging, repeated follow-up studies have shown that there has been no recurrence of obstruction.

**References**


**Comments by E. Douglas Wigle, M.D.*

Prior to answering the specific questions posed in relation to this 36-year-old woman with hypertrophic or muscular subaortic stenosis, attention should perhaps be drawn to the unusual features of this case. These are: (1) Patients with latent IHSS (obstruction to outflow only present with provocation) usually respond favorably to propranolol therapy. Rarely do their symptoms worsen with this therapy. (2) It is unusual for a patient with this condition to have a normal ECG and heart size, then four years later to have left ventricular hypertrophy and strain on ECG, plus marked cardiac enlargement on x-ray film.

The questions posed are entirely related to the future management of this patient. Should therapy be medical (\(\beta\)-adrenergic blocking agents) or surgical, and if the latter, what type of surgical procedure?

We usually give all patients with muscular subaortic stenosis a trial on propranolol therapy. In our own experience 100 percent of patients (10/10) with latent obstruction to outflow have had sustained symptomatic benefit, but only 36 percent of patients with resting obstruction to outflow (8/22) have benefited in the long term (for more than two to three years). A trial of propranolol could be given this woman, but we would not be optimistic that it would be of benefit since she responded adversely when the obstruction to outflow was only latent. Propranolol therapy rarely significantly alters the obstruction to outflow at rest, nor does it alter the echocardiographic abnormalities in the outflow tract, so we would not see any great benefit to be derived from objective testing (eg, echocardiography) to see if her outflow tract obstruction decreased. Whether this woman is given an initial (brief) trial of propranolol therapy or not, it would be our feeling that eventually surgery should be recommended, and that it should be performed in a center known to have successful surgical experience with this condition.

There are basically four surgical procedures that could be carried out: (1) ventriculotomy operation (without muscle resection);\(^1,2\) and (2) double myotomy, two incisions into the obstructing muscle of the outflow tract with resection of the intervening muscle\(^3\) (procedures 1 and 2 are carried out from an aortic approach); (3) using a combined aortic and left ventricular approach to carry out resection of the hypertrophic anterior end of the ventricular septum;\(^4,5\) and (4) mitral valve replacement.\(^6\) This latter type of operation is successful in relieving the outflow tract obstruction because the offending anterior mitral leaflet is removed (the systolic apposition of this leaflet with the ventricular septum is the cause of the obstruction to outflow).

Our own preference as to surgical procedure

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CHEST, VOL. 64, NO. 2, AUGUST, 1973
would be a simple ventriculomyotomy, in that it has been demonstrated to abolish the systolic anterior movement of the anterior mitral leaflet and thereby the obstruction to outflow and the invariable mitral regurgitation (when there was no independent mitral valve abnormality). The left ventricular end-diastolic pressure has also been reduced in the majority of instances and patients have had lasting symptomatic benefit (up to 11 years). The risks, as well as symptomatic and hemodynamic benefit to be derived from the first three types of surgical procedure (vide supra) are roughly comparable. We would prefer to reserve mitral valve replacement in this condition (with its attendant risks) for those patients with severe mitral regurgitation that can be demonstrated to be due to an independent mitral valve abnormality.

REFERENCES


