MEDICAL GRAND ROUNDS
PARKLAND MEMORIAL HOSPITAL
March 2, 1962

Atrial Septal Defect and Associated Anomalies

CASE #1: [redacted], 17 NM, [redacted]

The patient was seen in the allocation clinic with the complaints of difficulty in swallowing, and wheezing of 3 months' duration. His tonsils were enlarged and he had inspiratory musical rales over both lung fields. A routine electrocardiogram revealed an incomplete RBBB

From 1950 to 1952, the patient was admitted and treated in a Shreveport hospital for an active rheumatic carditis and recurrent upper respiratory tract infections. Since 1952, he moved to Dallas and led an active life.

Physical examination revealed a well-built young man. His blood pressure was 125/90. There were multiple inspiratory and expiratory wheezes. A grade 1-2 ejection murmur was heard over the 3rd L.I.S. The pulmonary second sound was accentuated, split and the splitting was fixed. The rest of the findings were essentially normal.

	Pressures	O2 Content, Vol.%
Pulmonary artery	35/20 (25)	14.16
Right ventricle	39/11	14.11
Right atrium	4	13.31
Superior vena cava	—	12.24
Left innominate vein	—	11.75
Brachial artery	—	15.94

Pulmonary blood flow	18.6 L/min.
Systemic blood flow	9.8 L/min.
Left-to-right shunt	8.8 L/min.

A 3 cm. diameter septum secundum defect was found at surgery. A well-formed rim of septum was present around the defect. The defect was successfully closed.

CASE #2: [redacted], 35 WF, [redacted]

At the age of 13, the patient developed a febrile episode and because her attending LMD heard a murmur, she was confined to bed for three months. The patient did not recall having had joint pains, nose bleeds or skin rash. She did fairly well until 3 years prior to admission, when she started experiencing general weakness and became easily fatigued. These symptoms became progressively worse during the few months prior to admission. She was referred to us with the diagnosis of mitral stenosis.

BP 120/86, pulse 68 and regular, chest symmetrical

Lungs were clear. The heart was slightly enlarged; a right ventricular heave was noted. An ejection systolic murmur was heard along the left sternal border (3rd-4th I.S.). The 2nd pulmonic sound was split both during inspiration and expiration. The rest of the physical exam was essentially normal.

	Pressure	O2%
Pulmonary artery	30/9 (15)	89
Right ventricle	32/2	88
Right atrium	(2)	87
Brachial artery	102/62	97
Superior vena cava	—	66
Innominate vein	—	63

Systemic blood flow 2.65 L/min.
Pulmonary blood flow 13.5 L/min.
Left-to-right shunt 10.85 L/min.

Phonocardiogram: A_2-P_2 = 0.06 sec. (inspiratory) and 0.04 sec. (expiratory)

A foramen ovale type of atrial septal defect was closed under direct vision (May 1960).

CASE #2: [redacted] 46 WM, [redacted]

At age 11, the patient had an illness that consisted of a fever, arthralgia and chest pain that was diagnosed "rheumatic fever". Since then, murmurs were found on examination. He was rejected from military service in 1942. He was asymptomatic until [redacted] 1961, when, following a period of stress, he had palpitations, became pale and had profuse sweating. Since then, he experienced exertional dyspnea, easy fatiguability and frequent episodes of palpitations.

BP 110/70, pulse 84/min. The lungs were clear. The heart was enlarged. A grade 4 systolic murmur, ejection in type, was heard all over the precordium, best along the L.S.B. Pulmonic second sound was loud and split.

	Pressure	O2 Saturation %
Pulmonary artery	69/22 (38)	—
Right ventricle	68/2	83
Right atrium	0	80
Superior vena cava	—	68
Innominate vein	—	54
Brachial artery	110/70	91

Systemic blood flow 2.3 L/min.
Pulmonic blood flow 11.3 L/min.
Left-to-right shunt 9.0 L/min.

The atrial septal defect was of secundum type, 4 cm. in diameter. The defect was successfully corrected on [redacted]/61.

CASE #4: [redacted], 56 WM, [redacted]

The patient is known to have had a "heart condition" all his life. 10 years ago he had a heart attack with subsequent exertional dyspnea. A year prior to admission, his physical activities became very restricted and he began noticing easy fatiguability, general weakness, and increased exertional dyspnea with the appearance of peripheral edema. The patient denied paroxysmal nocturnal dyspnea and orthopnea. He also denied hemoptysis, or pleuritic chest pain.

BP 110/60, Pulse irregular at 112/min. Respirations 16/min.

The neck veins were distended in a semi-recumbent position. The lungs were clear. Heart was enlarged. There was a harsh systolic ejection murmur best heard along the left sternal border, S2 in pulmonic area was greater than aortic second sound, split and fixed. There was no peripheral edema. Hemoglobin 17 gm. per 100 ml.

	Pressure	O2 Saturation %
Pulmonary artery	70/45 (54)	69
Right ventricle	59/14	—
Right atrium	(9)	high 69 low 91 (LA)
Brachial artery	110/60	90
Superior vena cava	—	43

Systemic blood flow 2.6 L/min.

Surgical correction of the atrial septal defect was thought to carry a high mortality rate. He subsequently developed pleuritic chest pain with blood-streaked sputum. The patient was placed on anticoagulants and maintained since ([redacted] 1961). He was seen [redacted]/62 in the clinic, doing fairly well.

CASE #5: [redacted], 31 NF, [redacted]

This patient was admitted to [redacted] on the morning of [redacted] 1961. She fell out of bed and was found to be unable to speak and had weakness of the right upper and lower extremities. She gave a history of hypertension for the past 3-4 years, untreated. A cerebral angiogram revealed occlusion of the left middle cerebral artery.

Physical examination revealed a right hemiplegia, blood pressureof 130/80. A systolic murmur (ejection) was heard over the left precordium and a thrill was felt along the L.S.B. (3rd-4th I.S.). The electrocardiogram revealed 1° A.V. block, right ventricular preponderance. Hemoglobin 12.1 gm., sickle cell trait.

	Pressure	O2 Saturation %	O2 Content
Pulmonary artery	24/13 (18)		13.60
Right ventricle	40/4		13.41
Right atrium	(4)		low 13.41; hi 11.76
Superior vena cava	—		11.10
Brachial artery	—	96%	15.87

Systemic blood flow	4.17 L/min.
Pulmonary blood flow	8.77 L/min.
Left-to-right shunt	4.60 L/min.

Retrograde ventriculography revealed no evidence of left ventricle to right atrial shunt.

TABLE I

INCIDENCE OF THE TYPES OF ATRIAL SEPTAL DEFECTS IN ADULTS

A. Abbott's Series (1000 cases)

400 had atrial septal defect

1. Pure: 73
   a. Foramen ovale.....40
   b. Cephalad part.....10
   c. Caudal part.....18
   d. Multiple areas.... 5
2. Associated with other anomalies: 327

B. Lewis, Winchell, Bashour (63 cases) - 1957

Types:
1. Foramen ovale..... 48 (76%)
2. High atrial septal defect.... 10 (16%)
3. Continuous defect..... 1/63
4. Low defects..... 3/63

C. Evans Bedford (180 cases) - 1960

Types:		Cases:
1. Foramen ovale		
a. Central.....	67.6%	
b. Inferior caval.....	26%	170
2. Superior caval.....	5.3%	
3. Total absence.....	1.1%	
4. Atrioventricular defect... (Ostium primum)	5.5%	10

TABLE II

ATRIAL SEPTAL DEFECT - HEMODYNAMIC (PRE- AND POSTOPERATIVE) FINDINGS

	Preoperative (38 patients)	Postoperative (20 patients)
Systemic Blood Flow	4.6 L/min.	6.1 L/min.
Pulmonary Blood Flow	17.1 L/min.	6.1 L/min.
Left-to-Right Shunt	12.6 L/min.	—
Pulmonary Artery Pressure range (12/4-82/37)*	40/16 mm.Hg	26/10 mm.Hg
PA mean (7-62)	24 mm.Hg	16 mm.Hg
In 6, systolic gradient (PV)	12-28 mm.Hg	< 5 mm.Hg
Pulmonary Vascular Resistance range (33-948)**	160 dynes/cm/sec.	240 dynes (49/677)
Left Atrial Pressure*** (In 9, 2-20 mm.Hg)	8 mm.Hg	—
Right Ventricular Pressure Work	387 Kg.m/hr.	98 Kg.m/hr.

	Pre	Post
1.	53/16	75/30
2.	82/37	80/29
3.	72/34	28/4

* 2 of the 8 had coexisting mitral valve disease

** The 3 highest (332, 543, 948) in associated mitral valve disease

*** In 3, no gradient across the defect was found; in the remaining 6 cases, it ranged from 1 to 10 mm.Hg

TABLE III

DIFFERENTIATION OF 4 TYPES OF I.A. COMMUNICATIONS

1. Ostium primum
2. Ostium secundum
3. Common atrium
4. Total anomalous pulmonary venous drainage

All have in common:

a. Septal murmur at pulmonary area
b. Wide splitting of P2

Clinical Differences:

a. Secundum and primum generally tend to develop normally, have no difficulties in childhood and show no cyanosis
b. Common atrium and total anomalous pulmonary venous drainage show early signs retarded development and intermittent or persistent cyanosis

LAD + RBBB = septum primum and common atrium
RAD + RBBB = septum secundum and total anomalous venous drainage

TABLE IV

SURGICAL INDICATIONS

A. Factors Influencing Operative Mortality in Atrial Septal Defect

	Cases	% Mortality
Congestive Heart Failure	18	39
Right Atrial Pressure (Peak)		
15 mm.Hg or more	10	50
< 15 mm.Hg	109	8
Right-to-Left Shunt		
10% or more	14	50
< 10%	105	7
Pulmonary Hypertension		
Systolic 75 mm.Hg or more	19	53
< 75 mm.Hg	100	4
Ratio of Total Pulmonary Resistance to Systemic		
0.5 or more	7	71
< 0.5	38	18

[The authors (McGoon, et al.) do not believe that the age factor is important but strongly suggest that surgery be done in the young adult as they tolerate surgery better even when pulmonary hypertension may be present.]

B. Relation of Mortality Rate to the Number of Factors

	Cases	% Mortality
None	80	1.3
1	22	4.5
2	9	56
3	6	83
4	—	—
5	2	100

TABLE V

SURGICAL RESULTS IN PRIMUM DEFECTS

1. Mayo Series (Kirklin): 66 patients

48 partial form (16 no symptoms, 3 marked symptoms)
18 complete form (11 severe symptoms)

Pulmonary hypertension: 16/48 with partial form
17/18 with complete form
(> 70 mm. in 13)

Results:

Partial: 4.2% in hospital
2 late deaths from A.V. block
2 others have persistent A.V. block

Complete: 12/18 died postoperatively (66.7%)

Causes of Death: Heart block, pulmonary hypertension, severe AV incompetence

2. Cooley Series

29 partial form - mortality 13%
(8 complete A.V. block)

24 complete form - mortality 64%

BIBLIOGRAPHY

Embryological Development

1. Edwards, J. Congenital malformations of the heart and great vessels. In: Pathology of the Heart (S.E. Gould, Editor), Thomas, Springfield, 1953.

2. Taussig, H. Congenital malformations of the heart. Commonwealth Fund, N.Y., 1947.

Surgical Anatomy of Atrial Septal Defects

3. Lewis, J., Taufic, M., Varco, R., and Niazi, S. Surgical anatomy of atrial septal defect: Experiences with repair under direct vision. Ann. Surg. 142:401, 1955 (Sept.).

4. Lewis, J., Winchell, P., and Bashour, F. Open repair of atrial septal defects. Results in sixty-three patients. J.A.M.A. 165:922, 1957 (Oct.).

5. Evan Bedford, D. The anatomical types of atrial septal defect - Their incidence and clinical diagnosis. Am. J. Cardiol. 3:568, 1960 (Sept.).

6. MacKrell, J., and Ibanez, R. Atrial septal defects - A clinicopathologic appraisal. Am. J. Cardiol. 2:665, 1958 (Dec.).

7. Kirklin, J., Wood, E., Burchell, H., and Edwards, J. Anatomic, physiologic and surgical considerations in repair of interatrial communications in man. J. Thor. Surg. 29:37, 1955 (Jan.).

Clinical and Physiologic Features of Atrial Septal Defects

8. Chapman, C., and Frazer, R. Clinical and hemodynamic features of uncomplicated interatrial defects in adults. Am. Heart J. 46:352, 1953.

9. Bashour, F., and Winchell, P. Clinical and physiologic studies before and after closure of atrial septal defects. Circulation 14:909, 1956.

10. Winchell, P., and Bashour, F. Some physiologic features of atrial septal defect. Am. J. Cardiol. 2:687, 1958.

11. Barber, J., Magidson, O., and Wood, P. Atrial septal defect. Special reference to the electrocardiogram, the pulmonary artery pressure and the second heart sound. Brit. Heart J. 12:277, 1950.

12. Leatham, A., and Gray, I. Auscultatory and phonocardiographic signs of atrial septal defect. Brit. Heart J. 18:193, 1956.

13. Leatham, A. Splitting of the first and second heart sounds. Lancet 2:607, 1954.

14. Boyer, S., and Chisholm, A. Physiologic splitting of the second heart sound. Circulation 18:1010, 1958.

15. Perloff, J., and Harvey, W. Mechanisms of fixed splitting of the second heart sound. Circulation 18:998, 1958.

16. Liu, C., and Jacono, A. Phonocardiography in atrial septal defect. External and intracardiac phonocardiograms. Am. J. Cardiol. 2:714, 1958 (Dec.).

17. Dimond, G., and Beuchimol, A. Phonocardiography in atrial septal defect. Correlation between hemodynamics and phonocardiographic findings. Am. Heart J. 58: 343, 1959 (Sept.).

Lutembacher's Syndrome

18. Lutembacher, R. De la stenose mitrale avec communication interauriculaire. Arch. mal. Coeur 9:237, 1916.

19. McGinn, S., and White, P. Interatrial septal defect associated with mitral stenosis. Am. Heart J. 9:1, 1933.

20. Roesler, H. Interatrial septal defect. Arch. Int. Med. 54:339, 1934.

21. Nadas, A., and Alimurung, M. Apical diastolic murmurs in congenital heart disease: The rarity of Lutembacher's syndrome. Am. Heart J. 43:691, 1952.

22. Ferencz, C., Johnston, A., and Wiglesworth, F. Congenital mitral stenosis. Circulation 9:161, 1954.

23. Bashour, F., and Simmons, D. Atrial septal defect with mitral valvulitis. Clinical and catheterization diagnosis. Ann. Int. Med. 48:1194, 1958.

24. Sambhi, M., and Zimmerman, H. Pathologic physiology of Lutembacher syndrome. Am. J. Cardiol. 2:681, 1958.

Electrographic-Vectorcardiographic Findings in Atrial Septal Defect

25. Toscano-Barboza, E., Brandenburg, R., and Swan, J. Atrial septal defect. The electrocardiogram and its hemodynamic correlation in 100 proved cases. Am. J. Cardiol. 2:698, 1958.

26. Blount, G., Munyan, A., and Hoffman, M. Hypertrophy of the right ventricular outflow tract. A concept of the electrocardiographic findings in atrial septal defect. Am. J. Med. 22:784, 1957.

27. Blount, G., Pryor, R., and Woodwark, G. The electrocardiogram in atrial septal defect. Heart Bulletin 8:55, 1959.

28. Davies, H., Pryor, R., and Blount, G. Electrocardiographic changes in atrial septal defect following surgical correction. Brit. Heart J. 22:274, 1960.

29. DeOliveira, J., and Zimmerman, H. The electrocardiogram in interatrial septal defects. Am. J. Cardiol. 2:694, 1958.

30. Burch, G., and DePasquale, N. The electrocardiogram and ventricular gradient in atrial septal defect. Am. Heart J. 58:190, 1959.

31. Tapia, F., and Proudfit, W. Secondary R waves in right precordial leads in normal persons and in patients with cardiac disease. Circulation 21:28, 1960.

Ostium Primum Syndrome

32. Rogers, H., and Edwards, J. Incomplete division of the atrioventricular canal with patent interatrial foramen primum (persistent common atrioventricular ostium). Am. Heart J. 36:28, 1948.

33. Kiely, B., Adams, P., Anderson, R., and Lester, R. T e ostium primum syndrome. AMA J. Dis. Child. 96:381, 1958.

34. Cahen, P., et al. Faux Syndrome de Lutembacher par persistence de l'ostium primum avec scission de la valve mitrale interne (Complexe de Rokitansky - Maud Abbott). Arch. mal. Coeur 45:203, 1952.

35. Sellors, T., and Somerville, W. The persistent ostium primum atrial septal defect (partial persistent common atrioventricular canal). Postgrad. Med. J. 37: 646, 1961.

36. Beregovich, J., Bleifer, S., Donoso, E., and Grishman, A. The vectorcardiogram and electrocardiogram in persistent atrioventricular canal. Circulation 21:60, 1960.

37. Burchell, H., DuShane, J., and Brandenburg, R. The electrocardiogram in patients with atrioventricular cushions defects (defects of the atrioventricular canal). Am. J. Cardiol. 4:575, 1960.

Surgical Indications and Results

38. McGoon, D., Swan, H., Brandenburg, R., Connally, D.,and Kirklin, J. Atrial septal defect: Factors affecting surgical mortality rate. Circulation 19:195, 1959.

39. Swan, H., Kortz, A., Davies, H., and Blount, G. Atrial septal defect, secundum. An analysis of one hundred patients undergoing open surgical repair. J. Thor. Surg. 37:52, 1959.

40. Crafoord, C., and Senning, A. Persistent atrioventricular canal. Surgical experiences. Am. J. Cardiol. 3:618, 1960.

41. Cooley, D. Results of surgical treatment of atrial septal defects. Particular consideration of low defects including ostium primum and atrioventricular canal. Am. J. Cardiol. 4:605, 1960.