THE STRAIGHT BACK SYNDROME

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SYNOPSIS

Twenty asymptomatic subjects with innocent systolic murmurs due to the straight back syndrome were studied. The diagnoses of the referring doctor were atrial septal defect in 10, pulmonary stenosis in 4 and systolic murmur of unknown aetiology in 6 subjects. An ejection systolic murmur of varying intensity and loudest over the pulmonary area was heard in all subjects. In 4 subjects, an associated systolic thrill was also palpable.

The following radiologic findings in the chest Xray were found (1) straightening of the dorsal spine and marked reduction in the antero-posterior diameter (mean 8.4 cms) of the chest in all 20 subjects; (2) left-ward displacement of the heart in 5; (3) prominent pulmonary artery in 3 and (4) "Pancake" appearance of the heart in 2 subjects.

The electrocardiogram was completely normal in 19 subjects. In one, frequent ventricular extrasystoles were seen.

Right heart catheterization carried out in 8 subjects revealed essentially normal findings. Pulmonary function tests done in 16 subjects showed mild restrictive defects in 2. M Mode echocardiography done in 12 subjects showed normal findings and specifically no mitral valve prolapse.

The straight back syndrome is yet another important cause of pseudo heart disease. Careful clinical examination, electrocardiographic and radiologic analysis nearly always enable a correct diagnosis to be made. Of crucial importance is the need to recognise the clinical signs that are produced by an uncomplicated loss of normal thoracic vertebral curvature, so as to obviate the misdiagnosis of organic cardiac disease, where none actually exist.

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INTRODUCTION

Cardiac murmurs simulating organic heart disease due to deformities of the thoracic cage such as pectus excavatum and scoliosis have been well documented and recognised in the past. However, the importance of a loss of a normal dorsal kyphosis of the spine as a cause of innocent systolic murmurs has not been adequately appreciated, although this was first pointed out by Rawlings as early as 1960 (1). This condition was appropriately termed by him as "the straight back syndrome" (SBS) and is an important cause of innocent systolic murmurs in the praecordium.

In this paper, we present the clinical, electrocardiographic and radiologic features of 20 subjects with the SBS. In addition, the lung function test results of 16, the haemodynamic data of 8, and the echocardiographic findings of 12 subjects are also described.

MATERIALS AND METHODS

A detailed clinical history and physical examination were carried out in all the 20 subjects. In the physical examination, particular attention was focussed on the spine, the skeletal configuration of the chest and whether there was any clinical evidence of Marfan's Syndrome. The diagnosis of a SBS was made only if a straightening of the spine was seen on clinical examination and confirmed radiologically.

In each subject, a resting electrocardiogram and at least one postero-anterior and one lateral chest Xray film was taken. The antero-posterior (A-P) diameters were measured from the level of the anterior border of the body of the eighth thoracic vertebra to the posterior border of the sternum. The trans-thoracic diameters were measured at the level of the diaphragms. The ratio of the A-P to the trans-thoracic diameter was then derived.

Similar measurements were obtained from 20 male subjects without any skeletal abnormalities and specifically without straightening of their spines, to serve as controls.

Pulmonary function tests were performed in 16 subjects who were seated and at rest. Gas volumes were expressed at body temperature and pressure saturated with water vapour (BTPS).

The forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) were measured in triplicate on a 9-litre Godart closed circuit spirometer. In each test, the highest value achieved was taken as the final result.

The total lung capacity (TLC) and its subdivisions were determined in duplicate using the closed circuit helium dilution technique. Functional residual capacity (FRC) was calculated by standard methods. An agreement of 200 ml or less was required of two successive determinations. Predicted normal values were calculated from regression equations based on normal data obtained in this laboratory (2).

Right heart catheterization was done using standard techniques and measurements in 8 subjects.

In 12 subjects, M Mode echocardiography was performed using a Picker 80C Echoview echocardiographic machine, a 2.25 megaH focused transducer, and a Honeywell strip chart recorder employing standard techniques (3), with the subject lying supine and turned slightly towards the left. Special care was taken to hold the transducer perpendicular to the chest wall while recording the mitral valve echoes so as to avoid false positive or false negative findings for mitral valve prolapse.

RESULTS

The ages of the subjects ranged from 16 to 35 years with a mean of 17 years. There were 18 males and 2 females. Nineteen of the subjects were Chinese, the last being a Malay. (Table I).

TABLE I
BIODATA OF 20 SUBJECTS

Age : 16 to 35 years (mean 17 years)

Sex : Male = 18; Female = 2

Ethnic group : Chinese = 19; Malay = 1

HISTORY

All the 20 subjects were asymptomatic and were referred for consultation because of a systolic murmur. A diagnosis of atrial septal defect was made in 10, pulmonary stenosis in 4, and systolic murmur of unknown cause in 6 subjects by the referring doctor. (Table II) Four subjects with systolic thrills over the pulmonary area were refused employment on the basis of a wrong diagnosis of organic heart disease by their previous doctors.

TABLE II
DIAGNOSIS OF REFERRING DOCTOR

	No of subjects
Atrial Septal Defect	10
Pulmonary Stenosis	4
Systolic murmur ? cause	6

PHYSICAL SIGNS

In all the 20 subjects, there was loss of the normal dorsal curvature of the spine. This was obvious on clinical examination with the subject sitting upright (Fig. 1) and was easily confirmed with a lateral Xray of the chest.

In none of the subjects was there any clinical evidence of Marfan's Syndrome.

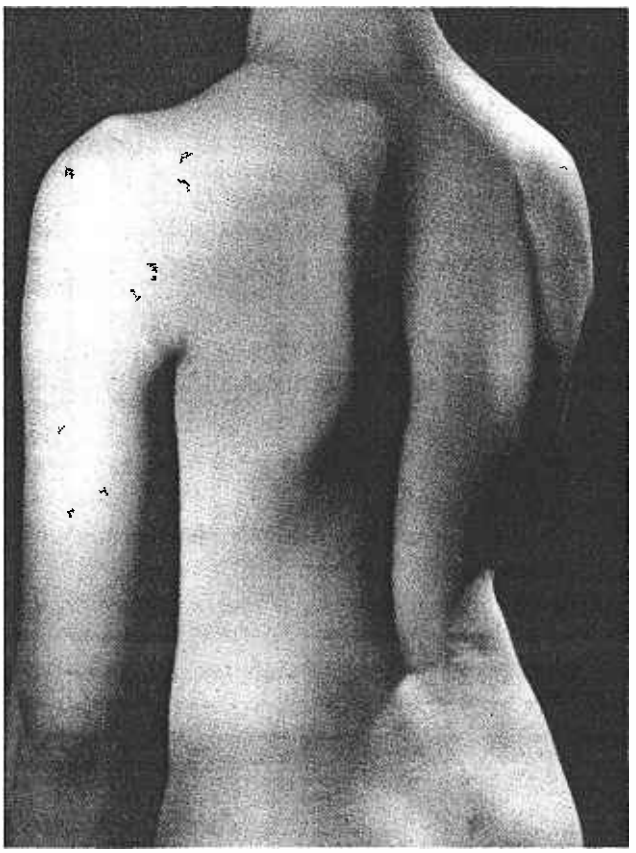


Figure 1: View of the posterior thorax. Note the loss of normal thoracic kyphosis and the straightening of the vertebral column between the two scapulae. (See text).

In 9 subjects, both pulmonary arterial pulsations in the second left intercostal space and left parasternal systolic pulsations over the third, fourth and fifth

intercostal spaces could be distinctly felt. Pulmonary arterial pulsations and left parasternal pulsations alone were felt in one and three subjects respectively. (Table III).

An early to mid ejection systolic murmur, maximum over the pulmonary area, varying in intensity from grade 1 to 4 (out of 6) was heard in all subjects (Fig. 2). In 4 subjects, these murmurs were associated with a systolic thrill. Two features of the systolic murmurs in the SBS were observed. First, there is a marked decrease in intensity of the murmur with deep inspiration which causes it to be hardly audible. Second, there is a marked increase in intensity of the murmur when the chest is compressed with the diaphragm of the stethoscope. Another characteristic feature of the murmur due to the SBS is a lack of radiation to the clavicles as is seen in the murmurs of aortic or pulmonary valvular stenosis. Clicks, diastolic murmurs, apical late systolic or holo systolic murmurs were not heard in this series.

The pulmonary second sound was assessed clinically in all subjects and was found to be widely split in inspiration in about half. In none however was there a "fixed splitting" of the second sound; but in about one quarter of the cases, the split was still heard in expiration.

RADIOLOGIC FINDINGS

As is seen in Table III, there was a marked reduction in both the antero-posterior (AP) diameter (mean 8.4 cms) and antero-posterior diameter/transthoracic (AP/TT) ratio % (mean 31.0%) as compared with the control series of subjects shown in Table IV. In the latter group of normal subjects, the mean AP diameter was 12.0 cms and the mean AP/TT ratio was 40.2%. It is noted that not only were the mean figures for the AP diameter as well as the AP/TT ratio in the SBS subjects lower than those of the normal subjects, but the highest figure in each range was also below the normal mean.

Three subjects showed minimal scoliosis in

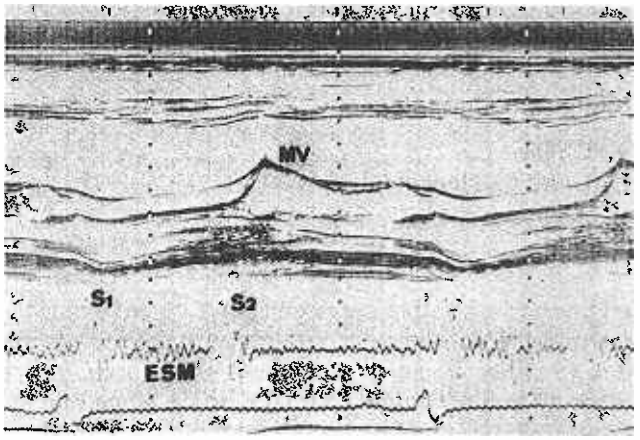


Figure 2: Simultaneous M Mode echocardiogram and phonocardiogram of Case 8. The echocardiogram shows a normal mitral valve. The phonocardiogram recorded at the pulmonary area shows an ejection systolic murmur and widely split second sound. S1 = first heart sound. S2 = second heart sound. ESM = ejection systolic murmur.

TABLE III

CASE NO	AGE & SEX	A-P DIAMETER (CM)	AP/TT RATIO (%)	C/T RATIO (%)	PALPABLE RV	PALPABLE PA	SYSTOLIC THRILL
1	19/M	7.1	26.7	40.2	+	+	+
2	18/M	6.2	26.7	47.4	+	+	+
3	19/M	10.1	27.2	30.7	+	0	0
4	19/M	6.4	27.6	50.0	+	0	0
5	17/M	7.7	29.3	41.8	+	+	+
6	17/M	7.4	29.3	45.9	+	0	0
7	18/M	8.1	30.2	34.4	0	0	0
8	18/M	8.9	34.1	39.5	0	0	0
9	18/M	10.4	35.6	45.7	0	0	0
10	17/M	9.2	35.7	48.9	+	+	0
11	16/M	9.1	35.7	39.7	+	+	0
12	17/M	8.9	35.8	48.1	+	+	0
13	18/M	9.0	36.0	39.7	0	0	0
14	17/M	8.9	36.0	48.6	0	0	0
15	17/M	9.2	36.1	47.8	0	0	0
16	21/M	9.1	36.2	40.6	0	+	0
17	35/M	8.8	36.3	54.3	0	0	0
18	21/M	8.9	36.4	37.6	+	+	+
19	15/F	7.4	28.4	51.0	+	+	0
20	22/F	8.8	36.8	45.0	+	+	0

A-P DIAMETER : Range - 6.2 to 10.4 cm (mean 8.4 cm) ± SE 0.3
AP/TT RATIO % : Range - 26.7 to 36.8 (mean 31.0) ± SE 0.9

AP = Antero-Posterior; TT = Transthoracic; PA = Pulmonary Artery;
CT = Cardiothoracic; RV = Right Ventricle

TABLE IV
RADIOLOGIC FINDINGS - 20 CONTROL MALE SUBJECTS

Antero-posterior diameter :
Range 9.8 to 14.2 cm (mean 12.0 cm)

Antero-posterior/transthoracic ratio % :
35.4 to 49.6 (mean 40.2)

TABLE V
RADIOLOGIC FINDINGS

	No of subjects
Straight thoracic spine	20
Scoliosis	3
Leftward displacement of heart	5
Prominent pulmonary artery	3
"Pancake" appearance of heart	2

addition to a straight spine and the leftward displacement of the cardiac silhoeutte seen in them could be due to either cause or a combination of both. In the other 17 subjects with a straight spine alone and no other skeletal abnormalities, a leftward displacement of the heart was seen in two, a prominent pulmonary artery in 3 and a "pancake" appearance of the heart in 2 subjects (Figs. 5-10).

3

HAEMODYNAMIC FINDINGS

Right heart catheterization was carried out in 8 subjects. The findings were completely normal except for 5 subjects with small insignificant systolic gradients (ranging from 4 to 11 mm Hg) in the right ventricular outflow tract. (Table VI).

PULMONARY FUNCTION TESTS

The dynamic and static lung volumes were measured in 16 subjects and the results are tabulated in Table VII. In only two subjects was a mild restrictive defect observed, where both the FEV, and FVC were less than 70% of the predicted normal values. In one of these two subjects (case 4) the TLC was below 70% of the predicted normal. All the other subjects had values within normal limits.

ELECTROCARDIOGRAPHIC FINDINGS

The ECG was normal in 19 out of the 20 subjects. One subject showed frequent ventricular extrasystoles. The mean frontal plane axis ranged from 0° to + 85°, with a mean of + 45°.

ECHOCARDIOGRAM

M Mode echocardiography was done in 12 subjects and showed completely normal findings. Specifically, none of the subjects had mitral valve prolapse.

DISCUSSION

In 1960, Rawlings (1) pointed out that a loss of the normal kyphosis of the thoracic spine could cause praecordial systolic murmurs which often closely

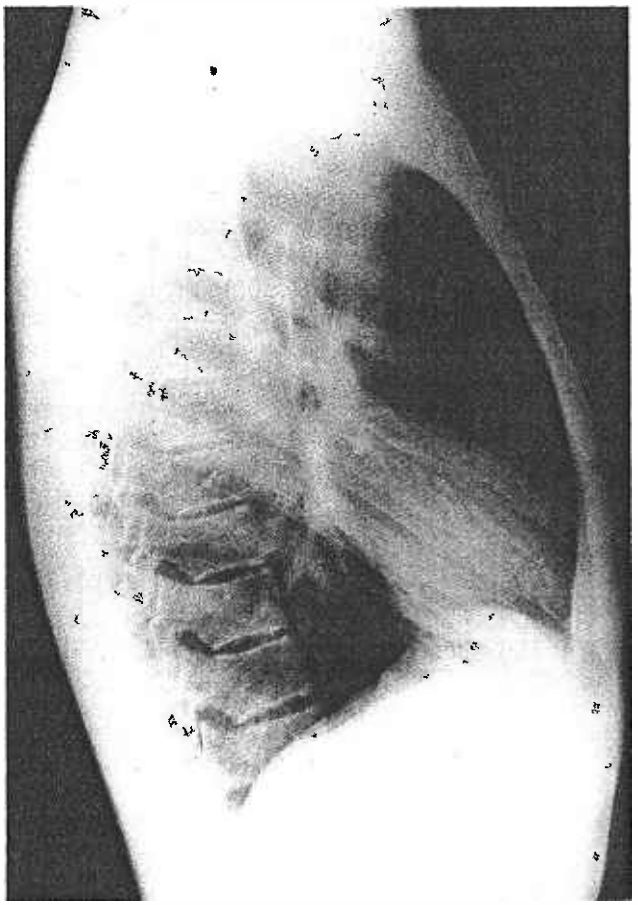


Figure 3

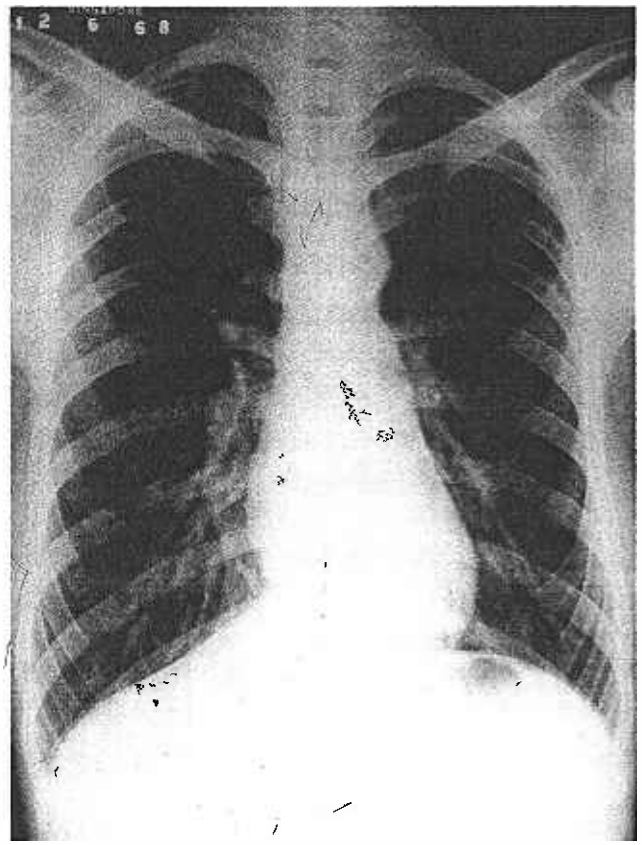


Figure 4

Figure 3 & 4: Lateral and P-A chest X-ray of a normal subject. Note the normal kyphosis of the thoracic spine. (See text).

TABLE VI
HAEMODYNAMIC DATA IN 8 SUBJECTS

Case No	Pressures in mm Hg					Cardiac Index L/min/m²
	RA (mean)	RV	PA		PCW (mean)	
4	6	28/5	28/10	(16)	10	2.6
6	5	32/6	24/12	(18)	11	4.8
10	4	34/4	25/10	(14)	10	4.1
12	6	27/5	23/12	(16)	9	3.8
17	5	26/5	26/9	(15)	8	3.9
18	5	33/3	22/12	(16)	8	3.0
19	5	30/5	22/10	(14)	9	3.2
20	5	22/3	22/8	(13)	7	4.0

RA = Right atrium; RV = Right Ventricle; PA = Pulmonary artery;
PCW = Pulmonary capillary wedge

TABLE VII
DYNAMIC AND STATIC LUNG VOLUMES (PERCENTAGE PREDICTED NORMAL) IN 16 SUBJECTS WITH STRAIGHT-BACK SYNDROME

CASE NO	LUNG VOLUMES PERCENTAGE PREDICTED NORMAL				
	FEV_1	FVC	FRC	RV	TLC
6	54.5	67.0	84.5	99.0	81.7
4	54.2	67.0	56.6	61.2	69.5
19	74.5	82.5	86.0	98.5	89.0
15	96.5	101.0	106.0	114.0	109.0
17	86.6	85.0	70.5	43.2	80.5
1	90.2	89.5	92.5	93.5	96.5
18	91.5	91.1	78.6	61.7	87.0
8	93.0	88.0	101.0	113.0	79.0
14	108.0	95.4	96.4	91.8	98.5
7	96.0	102.0	84.5	83.2	100.5
3	87.3	92.0	73.0	50.0	84.2
8	96.0	100.0	95.0	67.2	94.1
13	113.0	102.0	102.0	101.0	103.0
9	97.0	90.5	96.5	45.5	83.8
13	103.6	96.5	62.6	56.0	88.5
12	89.0	83.8	81.5	73.0	85.5
MEAN %	89.3	89.6	85.5	78.2	89.4
SE ±	4.2	2.7	3.6	6.0	2.6

FEV_1 = Forced expiratory volume in 1 second; FVC = Forced vital capacity;
FRC = Functional residual capacity; RV = Residual volume;
TLC = Total lung capacity

simulate organic heart diseases. This condition was appropriately termed by him as "the straight back syndrome" (SBS) and is an important cause of pseudo heart disease.

Absence of normal thoracic kyphosis (the straight back) is an asymptomatic skeletal defect and this osseous malformations appears to be congenital (5). This postulate is in accordance with the young ages of our patients which varied between 16 to 35 years (mean 17 years). With the straightening of the spine, the postero-anterior diameter of the chest is decreased, resulting in a forward and leftward displacement of the heart and great vessels. As a result of this, systolic murmurs occur in the second left intercostal space due to displacement of the pulmonary artery and to an exaggeration of the normal ejection vibrations due to the proximity of this vessel to the chest wall (6).

The typical murmur due to the SBS is a short ejection early to mid systolic murmur which is best heard at the pulmonary area.

In this series, this murmur varied in intensity from

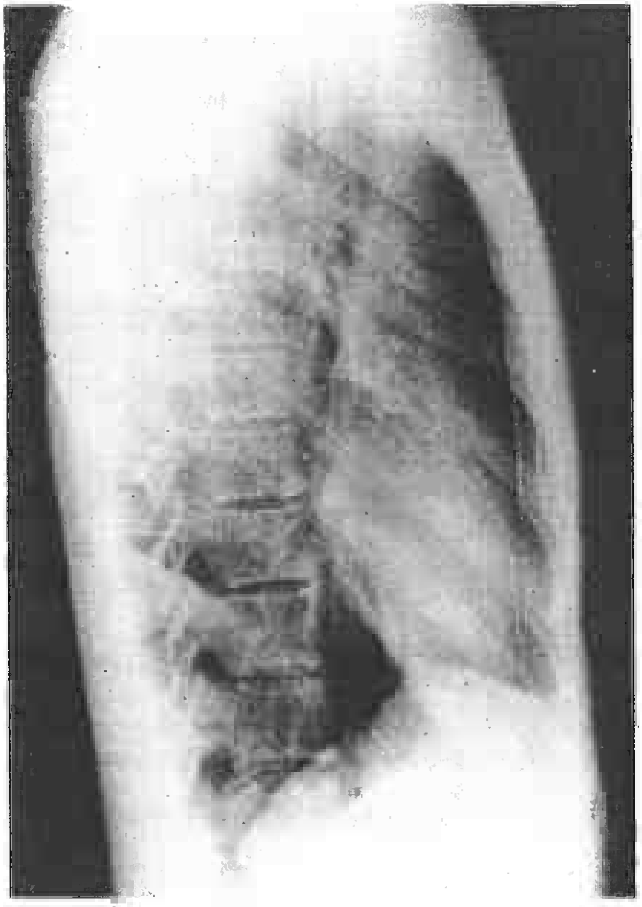


Figure 5

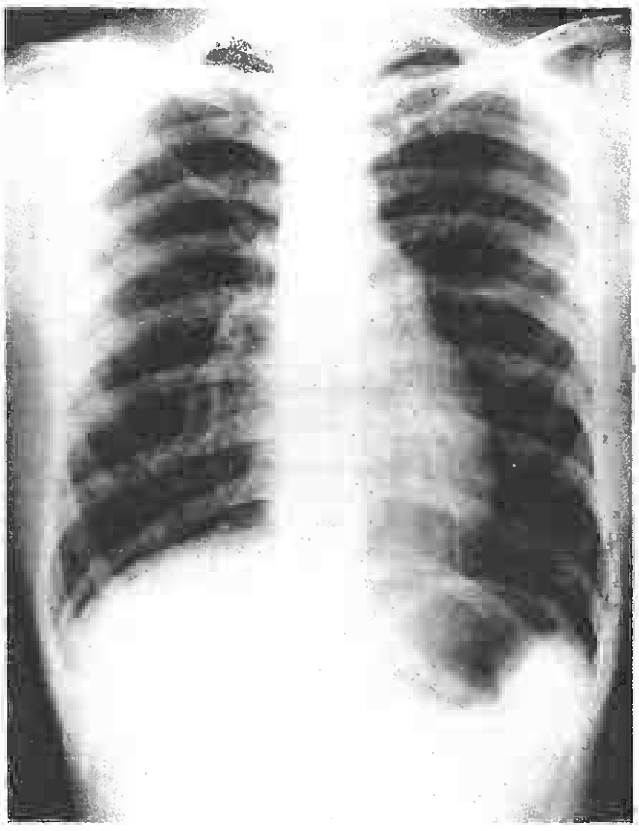
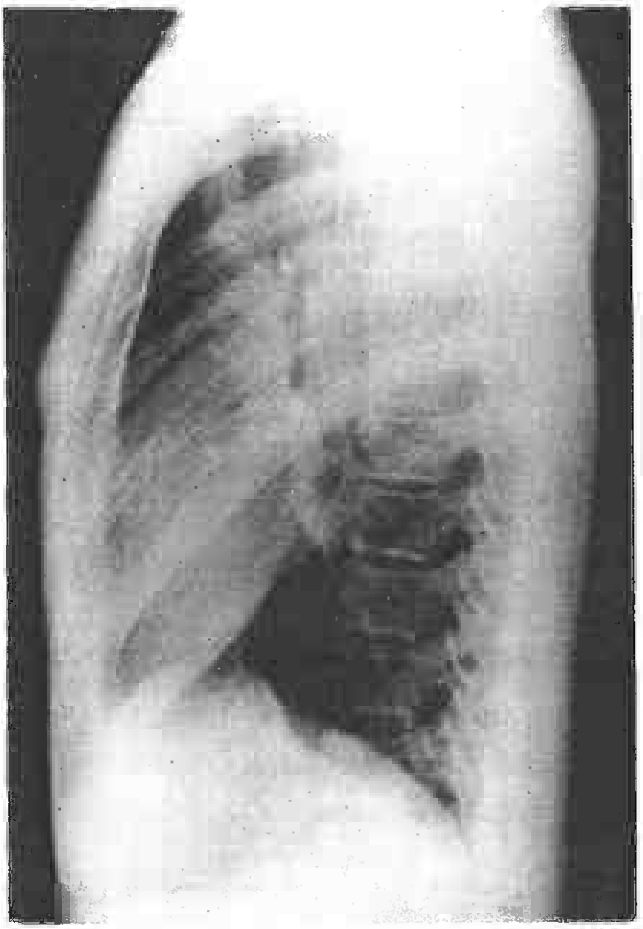


Figure 6

Figure 5 & 6: Lateral and P-A chest X-ray showing straight spine, reduced A-P diameter and prominent pulmonary artery. (See text).



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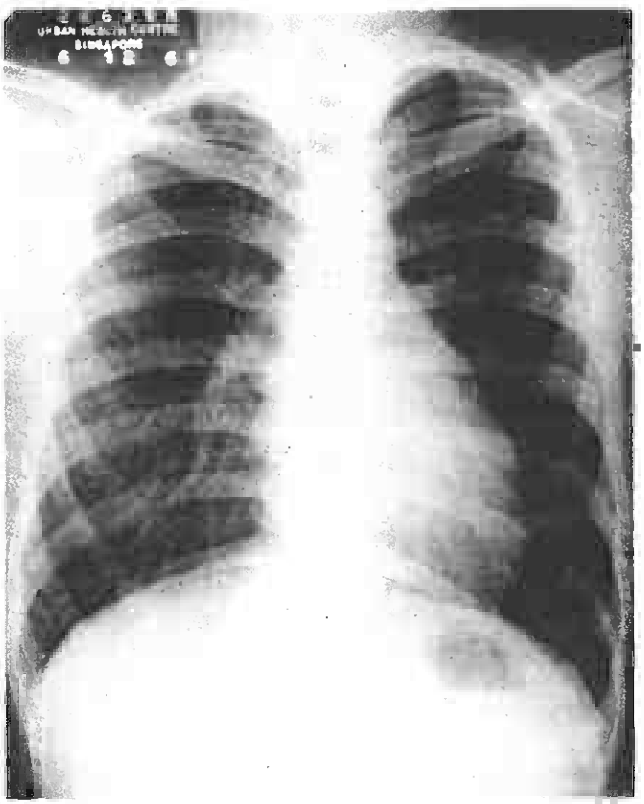

Figure 8

Figure 7 & 8: Lateral and P-A chest X-ray showing straight spine and left-ward displacement of the heart. (See text).

grade 1 to 4 (out of 6) and in 4 cases was so loud as to be associated with a systolic thrill. This latter finding has been described previously in the SBS (7) and is contrary to the popular belief that a systolic thrill always denotes an organic cardiac lesion. Two interesting features regarding the murmur were seen in this study. First, there was a marked decrease in intensity of the murmur with deep inspiration, which caused it to be hardly audible. Although most murmurs, except those due to right sided cardiac lesions, tend to decrease in intensity with inspiration, very seldom do they disappear completely, as frequently is the case with murmurs due to the SBS. Second, there was a marked and often dramatic increase in intensity of the murmur, when the chest wall was compressed with the diaphragm of the stethoscope. These two features strongly suggest that these murmurs are primarily due to a reduced antero-posterior diameter of the chest causing compression of the heart and great vessels.

The pulmonary second sound was accentuated in all cases simulating pulmonary hypertension and this again is due to the proximity of the pulmonary artery to the chest wall. In addition, this sound was widely split in half of our subjects, and in a quarter, this split was still heard in expiration. These findings could easily be confused with those of atrial septal defect.

Prominent pulsations in the second left intercostal space and in the praecordium were observed frequently in our subjects due to forward displacement of the heart. The latter finding can well be misdiagnosed as right ventricular hypertrophy if the diagnosis of SBS has not been realised.

The electrocardiogram in the SBS is usually normal. However an rSr pattern in lead VI, non specific ST-T wave changes and ventricular extrasystoles have all been reported previously (6).

The diagnosis of SBS is most conveniently made by just looking at the thoracic spine of the subject preferably in the sitting position (Fig. 1). The finding of a straight spine should be confirmed by a postero-anterior and lateral Xray of the chest. In our 20 subjects with the SBS, radiologic measurements revealed that there was a marked reduction in both the AP diameter as well as the AP/transthoracic ratio compared to the normal control subjects. As a result of this narrowing of the A-P dimension of the chest, certain characteristic findings in the chest Xray are seen. These include a leftward displacement of the cardiac silhoeutte, prominent pulmonary artery and the so called "pancake" appearance where the heart appears squashed and enlarged.

Haemodynamic findings in 8 subjects showed essentially normal results. Five subjects showed small insignificant systolic gradients across the right ventricular outflow tract ranging from 4 to 11 mm Hg. Such small gradients were also seen in 5 out of 9 cases reported by Matsuo et al (1973) (8).

Detailed lung function tests in 16 subjects showed mild restrictive defects in only 2 subjects. Hence in the great majority of instances, the straightening of the thoracic spine does not appear to impair pulmonary function to any significant extent.

M-mode echocardiography in 12 subjects showed normal findings and specifically no mitral valve

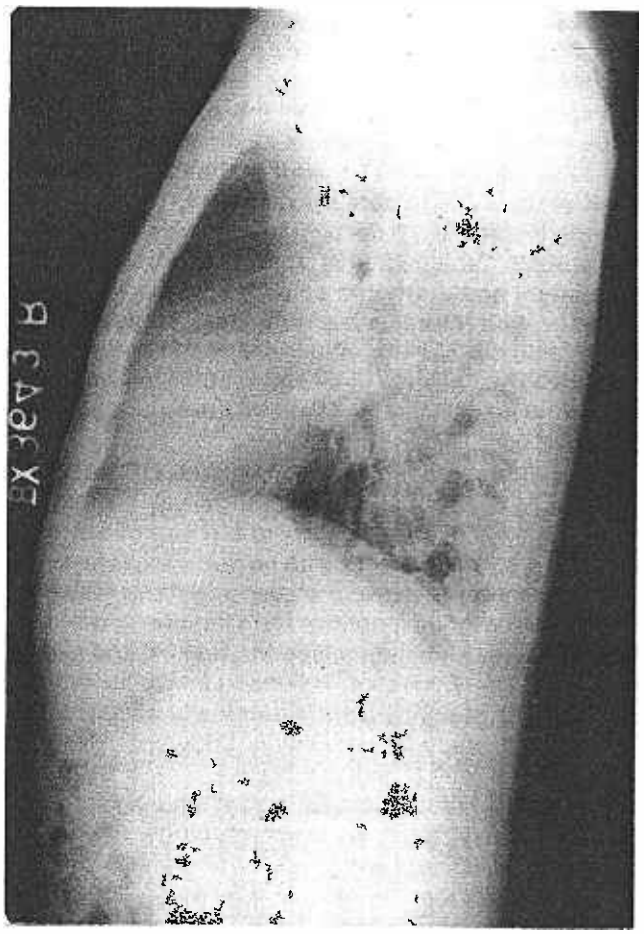

Figure 9

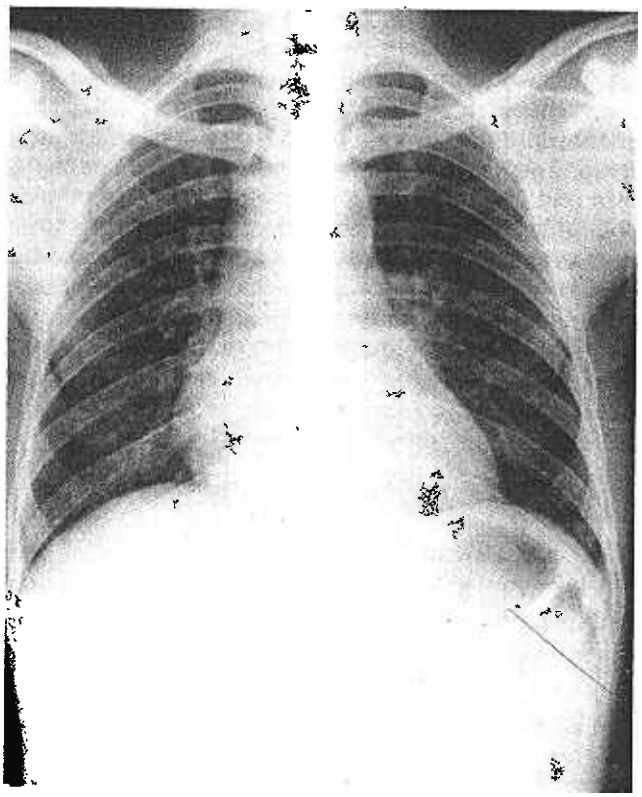

Figure 10

Figure 9 & 10: Lateral and P-A chest X-ray showing straight spine, cardiomegaly and "pan cake" appearance of the heart. (See text).

prolapse. It has been reported that thoracic skeletal abnormalities such as scoliosis, pectus excavatum and straightening of the thoracic spine are frequently encountered in patients with the prolapsing mitral value syndrome, being present in about 75% of cases in 1 study (9); sixty two per cent of the thoracic abnormalities were due to pectus excavatum, 17 per cent to a straight spine and 8 per cent to scoliosis. Saloman & co workers (9) concluded that since both pectus excavatum and a straight spine are very prevalent in patients with mitral valve prolapse, subjects with these thoracic skeletal abnormalities should not be diagnosed as having innocent systolic murmurs or pseudo heart disease until mitral valve prolapse has been excluded.

In addition, Udoshi and associates (10) recently reported a 54% incidence of mitral valve prolapse detected by echocardiography in a group of subjects with the SBS. In subjects with pectus excavatum, the incidence of mitral valve prolapse was 18%. However in this present study, none of the 12 subjects who had echocardiographic examination had mitral valve prolapse. The cause of the discrepancy in results between the study by Udoshi and his associates and the present study is not readily apparent. The true incidence of mitral valve prolapse in the SBS has obvious important clinical implications and clearly further studies need be done on large groups of unselected subjects with this syndrome to further clarify this problem.

The exact relationship, if any, between Marfan's syndrome, the SBS and the prolapsing mitral valve syndrome is a complicated but nevertheless intriguing one. In Marfan's syndrome, multiple thoracic skeletal abnormalities such as a straight spine, pectus excavatum and kypho-scoliosis are commonly present, in addition to the other classical features of the syndrome such as the "Marfan's habitus" and arachnodactyly. Furthermore, mitral valve prolapse is universally present in Marfan's syndrome (11). In patients with mitral valve prolapse but without overt features of Marfan's syndrome, thoracic skeletal abnormalities as has been described above are also frequently present (9). It has been suggested by some workers that mitral valve prolapse may indeed represent a form fruste of Marfan's syndrome (9) but evidence against this hypothesis has been well summarised by Devereux and his associates (12). Lastly, mitral valve prolapse has recently been reported by Udoshi and his associates to be present in about 54% of subjects with SBS (10) but was surprisingly not detected in any of the 20 patients examined echocardiographically in this present study. In the author's opinion, a straight spine may be a reasonably sensitive but non specific marker for mitral valve prolapse. This hypothesis however needs to be tested by further studies.

The clinical signs associated with the SBS can closely mimic organic heart disease. Indeed in this series, ten patients were referred to us with a diagnosis of atrial septal defect. However, a tricuspid "flow" murmur which is invariably seen in large atrial septal defects, is never seen in the SBS. Furthermore, in about ninety per cent of cases of atrial septal defect, right bundle branch block is seen and the presence of this electrocardiographic finding clearly favours this diagnosis. The systolic murmurs due to the SBS could well be mistaken also for either aortic or pulmonary valvular stenosis or ventricular septal defect. However, the systolic murmurs of pulmonary stenosis typically radiates to the left clavicle and is not localised to the pulmonary area as is seen in the murmur due to the SBS. The systolic murmur of ventricular septal defect can be differentiated by the fact that it is holo systolic in timing instead of ejection systolic and maximum over the lower left intercostal spaces instead of in the pulmonary area. Of crucial importance in the exclusion of these defects is the need to recognise the clinical signs that are produced by an uncomplicated loss of normal thoracic vertebral curvature.

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