Acta orthop. Scandinav. 39, 456–465, 1968

From the Orthopaedic Clinic, Karolinska Institutet, (Head: Prof. S. Friberg) and the National Clinic of Work Capacity Assessment (Head Dr. K.-D. Lundgren), Stockholm, Sweden.

LONG-TERM PROGNOSIS IN IDIOPATHIC SCOLIOSIS

By

ULF NILSONNE & KARL-DAVID LUNDGREN

Received 2.ii.68

The prognosis in idiopathic scoliosis has chiefly been discussed in terms of the possibility of forecasting the progress of a curvature observed at the onset of the disease. It is, of course, understandable that the orthopedic surgeon should concentrate on patients whose age makes them particularly susceptible to progression of the scoliosis and that most attention should then be paid to the opportunities for active therapy. At the same time, little is known about the fate of scoliotic patients after they have left the orthopedic surgeon's immediate field of interest. There are a number of general statements (*e.g. Kleinberg* 1951, *Margret Lange* 1962) to the effect that patients with severe scoliosis generally die before the age of 45–50, the deformation of the thorax impairing the heart-lung function and thereby resulting in an increased mortality. *Chapman et al.* (1939), in a group mainly composed of paralytic scolioses, even give a worse prognosis, with an average death age of 30 years. *Sulser* (1958) on the other hand reports, that in an autopsy material, the average death ages in kypho-scoliotic individuals were in agreement with those of the general population of Switzerland. The material was, however, rather mixed and even included osteoporotic changes of the spine in old patients. Thus it would seem that no systematic study has been published of the long-term prognosis in a pure group of idiopathic scoliosis.

The present study, on a group of patients with an observation time of up to 50 years, contains an analysis of the vital prognosis in idiopathic scoliosis and an account of the social and socio-medical function of these patients. A preliminary report has already been published (*Nilsonne & Lundgren* 1964).

MATERIAL

The study comprises a group of patients with idiopathic scoliosis who came to the Orthopaedic Clinic in Stockholm in the period 1/7 1913–30/6 1918, *i.e.* the first five years after the Clinic opened.

The concept of idiopathic scoliosis does not appear as a diagnosis in the case reports from that time. The earliest instance of this terminology that we were able to find in the literature dated from 1922 (*Kleinberg*). The Orthopaedic Clinic's records of diagnoses from the period covered by this study contain a variety of terms, including scoliosis rachitica, scoliosis congenita, scoliosis habitualis, scoliosis essentialis, and insufficientia vertebrae; moreover, these diagnoses are usually accompanied by a question mark to denote the examining physician's uncertainty about the etiology.

Some importance used to be attached to rickets as a cause of progressive scoliosis but this has been refuted by more recent investigations (*e.g.* Brunk 1951). Nearly all the present case reports include an ordinary full-length photograph of the patient, none of which display any external signs of deformities that might suggest rickets. It is thus most unlikely that any patient with rickets has been included in this investigation.

The available case reports contain very little, if any, information about the duration of the scoliosis and consequently the present cases may include an occasional instance of congenital scoliosis, *e.g.* due to vertebral malformation. This source of error cannot be very large, however, since the maximum incidence of congenital scoliosis is reported to be 5 per cent (*Kleinberg* 1951).

The photographs and, in some cases, X-ray pictures show that the other diagnostic terms exemplified above refer to structural, not functional scolioses.

The records from the period covered by this study include a large number of paralytic scolioses arising from polio. Since the sequelae of polio were a central orthopaedic problem at that time, we have accepted the diagnosis of paralytic scoliosis and such cases have been excluded.

Table 1. Distribution of patients by year of admission.

	1913	1914	1915	1916	1917	1918	Total
M	2	1	4	8	9	1	25
F	8	19	19	17	17	8	88
M + F	10	20	23	25	26	9	113

(In this and the following tables M = males, F = females.)

With these criteria, we have arrived at a group of patients whose diagnosis in modern terms would be idiopathic scoliosis. The patients either received no treatment or else they were prescribed physiotherapy and/or a supporting brace. No active attempts were made to correct the scoliosis by conservative or surgical means. From the modern point of view, the material may be regarded as a group of untreated patients with idiopathic scoliosis.

The composition of the material is presented in Table 1, which also shows the number of new cases per year during the period of the study. The total material comprises 113 patients, 88 female and 25 male. The female incidence is thus 78 per cent, which is in good agreement with the recognized sex difference in idiopathic scoliosis (*Lindeman* 1958), namely that rather more than three times as many females contract the disease as males.

Table 2. Distribution by age at admission.

Age at admission	M	F	M + F
7–9	4	6	10
10–14	4	36	40
15–19	11	33	44
20–24	4	7	11
25–29	1	4	5
30–34	1	2	3
Total	25	88	113

The distribution by age at the time of admission is given in Table 2. (The ages represent the difference between the year of admission and the year of birth). The mean age on admission for the total material is 15.9 years, while for the males it is 16.7 years and for the females 15.6 years (see also Table 5). These average ages seem to be higher than they are today, no doubt because the improved medical control of school children now results in an earlier orthopaedic assessment of any back trouble. If, instead, the mean age on admission is calculated only for those patients in whom the scoliosis had not yet become stationary, the result is 13.2 years for the females and 12.5 years for the males (assuming that skeletal growth and hence the progression of the scoliosis had ceased in women of 17 or more and in men of 18 or more, so that these cases are excluded). This indicates that a large proportion of the patients with idiopathic scoliosis in the period 1913–1918 were observed during the progressive stage in puberty, when they were roughly the same age as the patients encountered today.

VITAL STATISTICS

Inquiries were made to find out whether or not the patients were still alive at the time of this study. In the case of those who had died, the year and the cause of death were ascertained from the parish registry office. In most cases it was possible to obtain the case reports on these patients' last hospitalization and thereby confirm the reported cause of death. Further verification was obtained in many cases from the autopsy report. The patients have been followed up to the middle of 1963, which thus constitutes the end of the observation period for the living

individuals. The reported ages were calculated as the difference between the last year of observation (1963 or the year of death) and the year of birth. Eleven of the 113 patients could not be traced because the personal data in the records were too scanty to permit re-identification in the ordinary population register.

The distribution between living and dead patients is given in Table 3. Of all those who could be traced, 56 were alive and 46 had died at the time of this study, the corresponding figures for the 25 males being 10 alive and 12 dead (3 not traced) and for the 88 females, 46 alive and 34 dead (8 not traced).

Table 3. Distribution by "living", "dead" and "not followed".

	M	F	M + F
Living 1963	10	46	56
Dead	12	34	46
Not followed	3	8	11
Total	25	88	113

Table 4. Mean age of living and dead patients.

	Living 1963 No.	Mean age
M	10	60.0
F	46	62.6
M + F	56	62.1
	Dead	
	No.	**Mean age**
M	12	51.9
F	34	44.8
M + F	46	46.6

The mean ages of the living and the dead patients are given in Table 4. There is a considerable difference between these groups, the mean age for the former being 62.1 years and for the latter 46.6 years. In the dead group, the mean age of the women at the time of death, 44.8 years, is definitely lower than that for the men, 51.9 years.

The mean age of all the patients at the time of their first visit to the Clinic is given in Table 5. Those who have died were older at their first

visit than those who are still alive, which is to be expected statistically. The patients who could not be traced were in their turn older at their first visit than those who are known to have died. This probably means that many of the former died long ago, which would explain why they could not be traced.

The distribution of the dead patients by the year of death is given in Table 6. There is a strikingly low number of deaths during the early part of the period of observation, only 9 of the 46 traced individuals having died before 1941, 3 of them of Spanish influenza in 1918. This distribution provides further support for the hypothesis that many of those who could not be traced, died during the early part of the observation period.

The 11 cases which could not be traced were excluded from the calculations of mortality. If one accepts the argument that the majority of those who could not be traced should be included among the dead patients, their exclusion means that the mortality reported is an underestimation.

Table 5. Mean ages at year of admission.

	M	F	M + F
Living 1963	13.6	15.1	14.8
Dead	18.8	15.9	16.6
Not followed	18.7	17.9	18.1
Total	16.7	15.6	15.9

Table 6. Distribution by year of death.

Year of death	M	F	M + F
1918		3	3
1921–25		3	3
1926–30		1	1
1931–35		1	1
1936–40		1	1
1941–45	5	6	11
1946–50	4	5	9
1951–55		6	6
1956–60	2	7	9
1961–63	1	1	2
Total	12	34	46

Table 7. Number of observed (O) and expected (E) deaths by period and age.

	M		F		M + F	
	O	E	O	E	O	E
Period						
1913–20		0.6	3	2.1	3	2.7
1921–30		0.9	4	3.2	4	4.1
1931–40		0.9	2	2.7	2	3.6
1941–50	9	0.8	11	2.9	20	3.7
1951–60	2	1.1	13	3.9	15	5.0
1961–63	1	0.4	1	1.3	2	1.7
Age						
7–19		0.4	3	1.8	3	2.2
20–34		1.4	6	4.4	6	5.8
35–49	5	1.3	12	3.7	17	5.0
50–64	7	1.6	11	5.1	18	6.7
65–		0.0	2	1.1	2	1.1
7–44	2	2.6	13	8.6	15	11.2
45–	10	2.1	21	7.5	31	9.6
Total	12	4.7	34	16.1	46	20.8

Table 8. Mortality ratio (Quotient of O/E).

Age	M			F			M + F		
	O	E	O/E	O	E	O/E	O	E	O/E
7–44	2	2.6	0.8	13	8.6	1.5	15	11.2	1.3
45–	10	2.1	4.8	21	7.5	2.8	31	9.6	3.2
Total	12	4.7	2.5	34	16.1	2.1	46	20.8	2.2

Table 7 presents a comparison between the number of deaths observed and the number expected. The distribution was studied by decades as well as by age, the calculations being based on mortality tables for the Swedish population. It will be seen that the mortality is considerably higher than expected, particularly after 1940. There were thus 20 deaths in the period 1941–50 compared with an expected figure of only 3.7. As far as age is concerned, it is chiefly after the age of 45 that mortality is higher than expected: 31 actual deaths compared with 9.6 expected.

The relationship between observed and expected deaths can also be expressed as a mortality ratio (Table 8). This ratio is highest for the

group over 45 years of age. For the men in this group, the mortality ratio is extremely high, 4.8, but here it should be noted that the mortality for men is about 20 per cent higher in Stockholm than in the rest of Sweden (*Larsson* 1965); to some extent, therefore, this high figure may reflect the fact that 11 of the 25 males were resident in the city or county of Stockholm at the time of their first visit to the Clinic. The mortality ratio for all the patients aged 45 or more is 3.2, while that for the entire material is 2.2. In other words, the mortality in idiopathic scoliosis in this material is more than twice as high as in the general population.

Table 9. Distribution by cause of death.

Cause of death	Sex		Age at death	
	M	F	M	F
Pulmonary disease	1	11	53	18, 18, 19, 20, 24, 33, 39, 45, 45, 45, 54
Cardiac incompensation	7	9	39, 44, 47, 48, 51, 51, 54	27, 33, 38, 41, 46, 47, 48, 54, 56
Arteriosclerosis	3	6	62, 63, 64	55, 62, 64, 64, 67, 76
Tumour	1	4	47	33, 44, 45, 57
Other		4		48, 50, 50, 57
Total	12	34		

The causes of death are reported in Table 9. Lung disease comprises pulmonary tuberculosis and pneumonia. Cardiac incompensation refers to the patients who died in right heart failure. In several of these cases, the death certificate even mentions the term kyphoscoliotic heart. Under arteriosclerosis, the causes of death were cerebral haemorrhagia and cardiosclerosis. Only malignant tumors are included under tumor. Other causes of death include suicide and delirium.

Cardiac or pulmonary disease accounted for about 60 per cent of the deaths. The age at death is also given in Table 9, which shows that 13 of the 28 cases of heart or lung disease died before the age of 45.

SOCIAL AND SOCIOMEDICAL ASPECTS

Besides suffering cosmetically, patients with idiopathic scoliosis have a reduced physical work capacity, particularly in severe cases. The

marriage rate for the females in this material (excluding those not traced) has been taken as a measure of the social function. About 76 per cent are unmarried (Table 10). The proportion of married women is higher among the living patients, which might be expected since they have had more time in which to find a partner. Nevertheless, the figures show that an extremely large proportion of women with idiopathic scoliosis do not marry.

Table 10. Distribution of followed women by marital status.

F	No.	Unmarried No.	%
Living	46	31	67
Dead	34	30	88
Total	80	61	76

Table 11. Orthopaedic and disablement status for 50 living patients (see text).

Mean age living	62.1 years
Back symptoms	90 %
Disablement pension for scoliosis	30 %
Disabled but no pension	17 %

A questionnaire was distributed to all the living patients, asking about their work function and any subjective back trouble. Answers were received from 50 out of 56. Back symptoms were reported by 90 per cent (Table 12), mostly in the form of a feeling of tiredness or pains in the thoracic or lumbar spine on exertion. Many of the patients had used some type of corset most of their lives. Of those who answered, 30 per cent stated that they had received a disablement pension for their back trouble, while a further 17 per cent considered that they were unable to work and had apparently managed by living with their parents or relatives most of their lives. Altogether, therefore, 47 per cent of the living patients were disabled.

CONCLUSIONS

The study shows that there is an abnormally high mortality in idiopathic scoliosis. The average mortality among the 102 patients who could be traced is twice that in the normal population. The mortality appears to be highest after the age of 45. However, since the majority

of the 11 patients who could not be traced and who were therefore excluded from the mortality calculations probably died in the early part of the observation period, it is possible that the actual mortality before the age of 45 is higher than indicated by the present figures. Cardiac or pulmonary disease was the cause of death in 60 per cent of the patients who died.

Idiopathic scoliosis may also involve a social and sociomedical handicap. The cosmetic deformity is reflected in an extremely high proportion of unmarried women. Imbalance in the spinal column is evident from the markedly high incidence of subjective back trouble. An indication of the effect on the physical work capacity is provided by the finding that almost half of the living patients are unable to work.

These circumstances must be borne in mind when assessing the seriousness of a newly discovered case of idiopathic scoliosis.

SUMMARY

113 cases of idiopathic scoliosis (88 females, 25 males) first noted in the period 1913–18 were followed up to 1963. The material presents a marked hypermortality, with a mortality ratio of 2.2. Cardiac or pulmonary disease was the cause of death in 60 per cent of the individuals who died. The sociomedical effects of scoliosis are reflected in the very high proportion of the women who have not married as well as in the high percentage of the living individuals who are unable to work.

ACKNOWLEDGEMENT

Mr. *Tage Larsson*, M.D., F.L., Chairman of the Medical Research Council of the Swedish Life Offices, has given us kind and valuable advice in the statistical approach, for which we are most grateful.

RESUME

113 cas de scoliose idiopathique (88 femmes, 25 hommes) observés pour la 1ère fois entre 1913 et 1918 ont été suivis jusqu'en 1963.

L'échantillon présente une hypermortalité marquée, avec un rapport de mortalité de 2,2.

Les maladies cardiaques ou pulmonaires ont été les causes de la mort de 60 pour cent des individus décédés.

Les effets sociaux-médicaux de la scoliose se traduisent par une proportion élevée de femmes qui ne se sont pas mariées ainsi que par un haut pourcentage d'individus vivants incapables de travailler.

ZUSAMMENFASSUNG

113 Fälle von idiopatischer Skoliose (88 weibliche, 25 männliche) erstmalig registriert während der Periode 1913–1918, wurden bis zu 1963 beobachtet. Dieses Material zeigt eine markante Hypermortalität mit der Mortalitätsratio von 2.2. Herz- und Lungenkrankheiten waren die Todesursache in 60 Prozent der Todesfälle. Die Sozialmedicinische Bedeutung der Skoliose tritt in der hohen Zahl der unverheirateten Frauen sowie in der hohen Zahl der nicht arbeitsfähigen Individen hervor.

REFERENCES

Brunk, M. (1951) The importance of rickets in childhood as a cause of scoliosis in adult age. *Acta orthop. scand.* Suppl. 9.

Chapman, E. M., Dill, D. B. & Craybiel, A. (1939) The decrease in functional capacity of the lungs and heart resulting from deformities of the chest: pulmonocardiac failure. *Medicine* **18**, 167.

Kleinberg, S. (1922) The operative treatment of scoliosis. *Arch. Surg.* **5**, 631.

Kleinberg, S. (1951) Scoliosis. Willams and Wilkins Comp., Baltimore.

Lange, Margret (1962) Internistische Beobachtungen bei Skoliosen. *Verh. dtsch. orthop. Ges.* **50**, 47.

Larsson, T. (1965) Mortality in Sweden. *Suppl. Acta genet. et statist. med.* Vol 15.

Lindemann, K. (1958) Skoliosen. In Handbuch der Orthopädie. G. Thieme, Stuttgart.

Nilsonne, U. & Lundgren, K.-D. (1964) Långtidsprognosen vid idiopatisk scolios. *Nord. Med.* **72**, 1088.

Sulser, U. J. (1958) Zur Klinik und patologischen Anatomie der Kyphoscoliose mit besonderer Berücksichtigung der Lebenserwartung. *Cardiologia* **32**, 231.