Ankylosing spondylitis—experience with a self administered questionnaire: an analytical study

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SUMMARY A questionnaire drafted by the president of a self help group of patients suffering from ankylosing spondylitis (AS), the Danish Ankylosing Spondylitis Society (DASS), was completed by 179 of 184 (97%) consecutive patients with AS. The following results were found. The diagnosis of AS was delayed an average of 12.6 years for women and 9.5 years for men. No differences were found in age at onset of the disease. A comparison of juvenile and adult onset showed a higher incidence of initial peripheral articular manifestations in patients with juvenile ankylosing spondylitis. Stiffness progressed significantly in both sexes during the illness. Two men still had stiffness localised only to the lumbar spine after more than 20 years of illness. Twenty four of 47 women (51%) and 36/121 (30%) of the men had experienced extra-articular disease. Fifteen of 46 women (33%) and 18/121 (15%) of the men had iritis and conjunctivitis before the diagnosis of AS was established and later, in the course of the illness. The results of this study agreed with those of previous investigations. The advantage of using a questionnaire as a screening test is that many patients may be studied and information brought up to date in a simple and inexpensive way.

In the early 1970s 184 patients suffering from ankylosing spondylitis (AS) formed a self help group, the Danish Ankylosing Spondylitis Society (DASS). Since then the DASS has continued to grow and now has about 500 members.

The aims of the society are to extend knowledge and contribute to investigations on AS. To gain knowledge about the disease the DASS president, a non-physician, drafted a questionnaire about symptoms, treatment, social events, and heredity.

Every patient with AS who joined the society during the period 1972–1985 was invited to complete the questionnaire, and all did so. At this time the DASS had 184 members, 133 men (72%) and 51 women (28%).

The replies to the questionnaire have provided some important information about the epidemiology of AS in Denmark.

Materials and methods

The questionnaire addressed the following subjects: age of onset of initial symptoms related to the

disease, symptomatology, sites and progression of pain and stiffness, treatment, social events and heredity, and the delay in diagnosis.

Variables evaluated in this study were age at onset of disease, duration of disease, sites and type of initial symptoms which could be related to AS, development and progression of symptoms and their relation to time of day. The manifestations of extra-articular symptoms was also studied.

Completed questionnaires were received from all the 184 members of the DASS, but five were excluded because their diagnosis of AS had not been established by a rheumatologist, leaving 179 (97%) patients included. Initially, all had been examined and followed up through academic centres or by a rheumatologist. Later on, most of the patients were followed up by rheumatologists or general practitioners.

In some cases insufficient answers had to be excluded from the study, resulting in a varying number of total answers to various questions.

The duration of disease was defined as the period between onset of the first symptoms related to AS and the time when the questionnaire was completed.

Axial onset was defined as symptomatic disease of the cervical, thoracic, or lumbar spine. Peripheral articular onset included symptoms from all other

joints, and in mixed onset disease both peripheral and axial symptoms were present simultaneously.

Table 1 shows the original questions relating to pain and stiffness. To explore the extent of stiffness in relation to sex and duration of disease four different phases were described: phase 1—stiffness involving the lumbar spine; phase 2—involving the lumbar and the thoracic spine; phase 3—involving the whole spine; phase 4—involving the whole spine and the leg joints.

None of the patients was recalled for physical or laboratory investigations.

Statistical significance was calculated by the χ^2 test.

Results

One hundred and thirty one of 133 men (98%) and 48 of 51 women (94%) completed the questionnaire. The mean duration of the disease was 19.9 years in women (range 1–43) and 14.6 years in men (range 1–58).

Figure 1 shows the number of patients in different age groups at onset of symptoms which could be related to AS; 32/52 (62%) of the men and 11/19 (58%) of the women developed low back pain as their first symptom at an age of 21–25 years, 99% of the patients had symptoms before the age of 40. The age range for onset of illness was 10–41 years for men and 12–38 years for women (Fig. 1).

The average delay in diagnosis was 12.6 years for women and 9.5 years for men. Disease beginning before 16 years of age was considered as juvenile

Fig. 1 Age of the patients at onset of symptoms related to ankylosing spondylitis.

onset, and was seen in 21/179 (12%) of the patients (14 male, seven female).

Table 2 compares the location of initial symptoms in patients with juvenile and adult onset AS. Axial symptoms were the primary symptoms in most of the patients. Peripheral articular manifestations, however, occurred significantly more often as the first symptom in patients with juvenile onset ($p=0.005$).

Fifteen of 41 women (37%) and 40 of 117 men (34%) with adult onset developed peripheral articular involvement later during the course of illness. This did not occur in patients with juvenile onset if axial symptoms had developed.

Significantly more patients had symptoms in the legs than in the arms (Table 3).

Pain in the cervical spine was described by two of 41 women (5%) and nine of 117 men (8%) with adult onset of the disease. This was not found in patients with juvenile onset.

Of 160 patients, 108 (68%) experienced their

Table 1 Questions relating to pain and stiffness

1	When did you begin to experience pain which you believe was a result of Bechterew's disease—year?
2	Where did the pain start (sacrum, low back, shoulder, neck, knee, elsewhere)?
3	What has been the course of the pain (constant, steadily increasing, intermittent, decreased, or stopped)?
4	At what time of day did the pain occur when it initially appeared (early morning, mid-morning, noon, afternoon, evening, night)?
5	Is it currently constant or do you have pain free periods?
6	Are the pains on one side or on both sides?
7	What has been the course of the stiffness? First phase: the lumbar spine; second phase: the lumbar and the thoracic spine; third phase: the whole spine; fourth phase: the whole spine and the leg joints; other? Specify the time of debut of the stiffness in each phase.
8	Is the stiffness constant?

Table 2 Comparison of the location of initial symptoms between patients with juvenile and adult onset ankylosing spondylitis.

	Peripheral articular		Axial		Mixed	
	F	M	F	M	F	M
Juvenile onset (<16 years)	4/7	3/14	3/7	6/14	0	5/14
Adult onset (≥16 years)	1/39	14/115	31/39	80/115	7/39	21/115

Table 3 Symptoms in the arms and legs, according to sex, in patients with ankylosing spondylitis

	Women (n=15)	Men (n=40)	Total (n=55)
Arms	2	3	5
Legs	13	37	50

most severe pains in the evening, during the night, and in the morning. No other specific variation in the pains was described.

Table 4 shows that for the course of pain in relation to duration of illness there was no difference between men and women.

In the women stiffness always appeared in less than five years and was localised to the lumbar spine. Further progression in stiffness was first described only later. The men had stiffness involving the spine and joints in a more complex way from the very beginning.

Nine of 23 men (39%) had stiffness localised to the lumbar spine. Another nine of 23 men (39%) reported that stiffness was localised to the whole spine. Five men developed stiffness in the cervical spine before the lumbar and thoracic spine were involved. Two of 26 men (8%) reported that stiffness was still localised only to the lumbar spine after more than 20 years of illness.

The stiffness progressed significantly in both sexes during the illness ($p < 0.001$). There was no significant difference between the sexes in this.

Twenty four of 47 women (51%) and 36/121 (30%) of the men had experienced extra-articular symptoms.

In 15/46 (33%) of the women and 18/121 (15%) of the men iritis and conjunctivitis occurred before the diagnosis of AS was established as well as later during the course of the illness. Seven of 45 women (16%) and 14/122 (11%) of the men described

symptoms from the gastrointestinal tract, but this included patients with discomfort resulting from drug treatment as well as patients with inflammatory bowel disease—for example, ulcerative colitis or Crohn's disease.

Discussion

Ankylosing spondylitis is a common systemic disease with a prevalence near 1%.¹ The diagnosis is based upon clinical criteria alone or together with radiological evidence of bilateral sacroiliitis.^{2,3}

In all the cases included in this study the diagnosis of AS had been established by a hospital specialist or rheumatologist. We accepted the validity of this information without retesting. In a study of Calin *et al.* the diagnosis of AS was evaluated and validated in 91–96% of their cases.⁴

Our questionnaire was designed from a layman's point of view. The clinical course of AS in this study, therefore, is based on symptoms which patients find of interest.

We compared our results with those of previous well-reputed studies and found that a self-administered questionnaire was effective and as a screening test had the advantage of facilitating the study of many more patients and bringing information up to date in an inexpensive and simple way.

AS was previously considered a predominantly male disease, but in recent years studies indicate no differences between the sexes.⁵ In this study the male to female ratio was 2.7:1, which accords with other results.^{4,6,7} Whether or not the ratio in our study reflects the true ratio in the population is open to question as our patients were a selected group.

These patients might have had a more active disease than patients with self limiting symptoms, and the male dominance in our study might be explained by the fact that women often have a somewhat milder disease or perhaps are less commonly diagnosed. On the other hand, women may be more likely to join a self help society.

As this study includes a comparatively small number of patients we are not able to comment on the trend towards increasing age at onset described by Calin *et al.*⁴ Ninety nine per cent of our patients developed symptoms which could be related to AS before the age of 40, and later age at onset was common. In accordance with the results of Gran Østensen, and Husby,⁵ we found no difference between the age at onset of disease in men and women.

A delay in the diagnosis of AS was expected.⁸ The men had the shortest delay, including those with juvenile AS. Our results are an average of the whole period 1972–1985. In recent years the delay is

Table 4 Course of pain in relation to duration of illness

Course of pain	Duration of illness (years)					
	<15		≥15 <30		≥30	
	F	M	F	M	F	M
Intermittent	5	21	3	11	1	0
Steadily increasing	4	34	7	26	6	4
Constant	3	8	2	11	0	2
Decreased/stopped	0	2	4	5	0	1
Total number	12	65	16	53	7	7

diagnosis for both women and men seems to have decreased.⁴ The reasons for this are not clear. It may be that more attention has been paid to AS.

Differences between juvenile and adult onset of AS were found in previous studies.⁹⁻¹¹ We confirmed that peripheral articular manifestations were significantly more common as initial symptoms in patients with juvenile AS. Furthermore, none of our patients with juvenile AS developed peripheral arthropathy after the onset of back symptoms.

Ladd *et al* found cervical spine involvement in patients with juvenile AS only rarely and considered this clinical feature to be of value in distinguishing these patients from those with juvenile rheumatoid arthritis.⁹ In agreement with these findings none of our patients with juvenile AS had pain involving the cervical spine.

In this study the patients experienced their most severe pain during the evening, night, and morning. However, what does the patient experience as stiffness? Pain or restriction in range of movement, or both, are described as stiffness by patients in other investigations.¹² This is perhaps not surprising, but as morning stiffness is a characteristic feature of AS^{13 14} we have to be careful when using this symptom as a clinical feature.

Carette *et al* suggest that a predictable pattern of AS emerges within the first 10 years after onset and that AS can have a benign course.¹⁵ The present cross-sectional study does not predict the course of AS, but some patients described relief of AS after more than 10-15 years of the illness.

Studying the course of stiffness, we found a significant correlation between progression of stiffness and duration of illness. Although there was a trend for stiffness to run a more progressive course in men than in women, at least during the first five years of illness, the number of women was too small to allow statistical analysis. A few men reported that stiffness was limited to the lumbar spine even after more than 20 years of illness.

It is usually considered that AS may be completely arrested at any stage. The results for pain and stiffness suggest that this is exceptional.

Except for the eye involvement, the incidence of extra-articular manifestations was remarkably low, even in patients with considerable disease duration.

This information is of great value to the prognosis for the individual patient. A clinical examination might have shown more patients with extra-articular disease. It does not make sense, however, to detect a group of patients who find their extra-articular symptoms not worth mentioning.

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