Beliefs about medications: a questionnaire survey of people with rheumatoid arthritis

R. Neame and A. Hammond

Objective. To investigate beliefs about medications held by people with rheumatoid arthritis (RA), what factors are related to these specific medication beliefs, and whether these beliefs influence adherence.

Methods. The design was a cross-sectional postal questionnaire survey of people with RA. The Beliefs about Medicines Questionnaire was used to assess beliefs about the necessity of medication and concerns about it. Questionnaires were mailed to 600 out-patients with RA.

Results. The response rate was 57.3%. Most (74.3%) respondents agreed or strongly agreed that their arthritis medications are necessary for their health. However, 47.4% were concerned about potential adverse consequences. The overall necessity score (mean 19.2, s.d. 3.13) was higher than the concerns score (mean 15.84, s.d. 3.53; P < 0.001). Greater disability was associated with higher necessity scores (r = 0.36; P < 0.001). Greater helplessness correlated with higher concerns scores (r = 0.49; P < 0.001). There was no association between RA knowledge and beliefs about medications (necessity scale, r = 0.02, P = 0.66; concerns scale, r = -0.08, P = 0.14). Concerns scores for non-adherent participants (mean 17.88, s.d. 3.29) were higher than for the adherent group (mean 15.64, s.d. 3.51; P = 0.002).

Conclusions. Most people with RA have positive beliefs about the necessity of their medication. However, levels of concern are high and associate with helplessness and non-adherence. The Beliefs about Medicines Questionnaire may identify people at risk of poor adherence and provide a focus for patients to discuss their beliefs, providing opportunities to improve adherence.

Key words: Rheumatoid arthritis, Beliefs about medicines, Knowledge, Adherence.

Beliefs and attitudes about illness influence adherence to treatment [1, 2]. In fact, beliefs about medicines predict adherence more strongly than sociodemographic or clinical factors [2]. It is estimated that 30–50% of people do not adhere to their prescribed treatment [3, 4]. Non-adherence may result in unnecessary health costs, investigations, changes in treatment, morbidity and mortality [5]. Investigating medication beliefs is especially important in rheumatoid arthritis (RA) as it is a chronic disease, and people are usually advised to take potentially toxic drugs, including non-steroidal anti-inflammatory drugs (NSAIDs) and disease-modifying anti-rheumatic drugs (DMARDs), which may be only partially effective. Being able to identify people with RA at risk of medication non-adherence could assist in the design and appropriately timed delivery of interventions to increase adherence, thus improving health status and reducing costs.

There is a relative paucity of information regarding the medication beliefs held by people with RA. Two recent studies have investigated these. Berry et al. [6] surveyed both new (n = 37) and existing (n = 44) rheumatology clinic patients receiving medication for ‘painful musculoskeletal conditions’ (potentially a third of whom had RA). New patients were mainly taking NSAIDs and existing patients mainly DMARDs. Perceived benefits of medication were primarily reduction of pain, stiffness and swelling, and perceived risks were primarily side-effects and becoming dependent on drugs. Although existing patients perceived their medications to be significantly more risky than did new patients, these concerns were still only at modest levels. A qualitative study (n = 29) with people with RA identified that most believed they had no choice about whether or not to take DMARDs as these were considered necessary to preserve joints, reduce pain and improve quality of life [7]. Participants expressed strong concerns about taking their medications, particularly with regard to potential toxicity and long-term effects. This study emphasized the tension experienced by people with RA with respect to the importance of their medication and their concerns about it.

Social cognition theories of health behaviour (e.g. the Health Belief Model) suggest that four key elements determine an individual’s adherence to treatment: threat of the illness (perceived severity and susceptibility), positive outcome expectancy (perceived benefits from treatment), barriers to using the treatment (e.g. expected disadvantages of treatment) and intent (intention to adhere to the treatment regime) [1]. Self-regulatory theory also emphasizes that treatment perceptions and illness representations influence medication adherence [8]. These theories propose that people undertake a cost–benefit analysis, considering whether their beliefs about the necessity of medications for maintaining health outweigh their concerns about the potential adverse effects of taking them. The Beliefs About Medicines Questionnaire (BMQ) is a tool for evaluating people’s beliefs about medicines [9]. It has been validated for use in patients with chronic illnesses and has been shown to predict adherence to treatment amongst other groups, e.g. people with asthma. People with strong beliefs in the necessity of taking medication to maintain their health were found to be more adherent to treatment, and those with higher levels of concern about medication, commonly about the dangers of dependence and long-term side-effects, were more likely to be non-adherent [2].

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The aims of this study were to use the BMQ to investigate the beliefs about medications held by people with RA. We also investigated what factors are related to these specific medication beliefs, and whether these beliefs influence adherence to drug treatments.

**Methods**

**Questionnaire**

A self-report questionnaire was used to collect data on six key areas.

**Beliefs about medicines using the Beliefs about Medicines Questionnaire [9]**. The BMQ consists of two five-item scales assessing patients’ beliefs about the necessity of prescribed medication for controlling their disease and their concerns about potential adverse consequences of taking it. Respondents indicate their degree of agreement with each statement on a five-point Likert scale, ranging from 1=strongly disagree to 5=strongly agree. Scores obtained for individual items within both scales are summed. Thus, total scores for the Necessity and Concerns Scales range from 5 to 25. Higher scores indicate stronger beliefs. A necessity–concerns differential is calculated as the difference between the necessity and the concerns scales, with a possible range of −20 to +20. This differential can be thought of as the cost–benefit analysis for each patient, for whom costs (concerns) are weighed against their perceived benefits (necessity beliefs) [2].

**Sociodemographic factors.** These are age, gender, highest educational level achieved and current employment status.

**Disease features.** Self-reported duration of RA was recorded. Functional status was measured using components of the Multidimensional Health Assessment Questionnaire (MDHAQ) [10]. This includes the modified Health Assessment Questionnaire (mHAQ, score range 1–4), 100 mm visual analogue scales (VAS) for pain and fatigue, a psychological distress scale (consisting of four items: stress, anxiety, depression and sleep quality, score range 1–4) and a Rheumatology Attitudes Index (RAI, score range 5–40) [11–13]. The MDHAQ RAI consists of five items of the learned helplessness subscale [13], with three additional items about medicine attitudes. Higher scores indicate greater levels of helplessness.

**Drug adherence.** The item ‘I often do not take my medicines as directed’ within the RAI was used to indicate drug adherence. Participants responded on a five-point scale (strongly disagree to strongly agree). Responses were dichotomized into an adherent group (strongly disagree/disagree) and a non-adherent group (strongly agree/agree/neither agree nor disagree, i.e. not taking treatment as directed).

**DMARD experience.** This comprises the duration of DMARD therapy, the number of DMARDs taken to date, and recollection of any DMARD adverse effects (dichotomized into those who did and those who did not experience them).

**Knowledge of RA.** The Arthritis Knowledge Questionnaire RA-specific subscale Form B [14] was used. This consists of 11 items assessing general knowledge about RA and drug treatments. For each question, respondents select one correct answer from five possible answers, including a ‘don’t know’ option. A percentage score is calculated. In two drug-specific questions, ‘sulphasalazine’ was substituted for ‘hydroxychloroquine’, and ‘methotrexate’ for ‘azathioprine’, as these are now much more frequently prescribed.

**Ethical approval**

Ethical approval was obtained from the Southern Derbyshire Local Research Ethics Committee (LREC).

**Participants**

The questionnaire was piloted with 10 out-patients with RA and subsequently modified to improve clarity. The questionnaire was then mailed to adult out-patients with RA registered with our electronic DMARD-monitoring database. The database was sorted in alphabetical order by surname and the first 600 people were selected. Freepost envelopes were provided for return of the questionnaires. The LREC required all replies to be anonymous, and so no reminders to non-respondents could be sent.

**Data analysis**

Data were analysed using SPSS v10.1 [15]. Data distributions were normal and therefore parametric analysis was conducted. Bivariate correlations were used to examine the associations of concerns scores and necessity scores with the variables of interest (sociodemographic, disease-related, DMARD-related and knowledge of RA). Partial correlations were used to further examine these associations with the remaining variables held constant. For nominal variables (e.g. gender, DMARD adverse effects, adherence group), two-tailed independent t-tests were used.

**Results**

**Response rate**

Three hundred and forty-four of 600 questionnaires were returned, a response rate of 57.3%. Some respondents did not complete every questionnaire item. Non-respondents were similar to the total sample mailed in terms of age and gender.

**Sample characteristics**

Around two-thirds (220/328 or 67%) of respondents were women, and 172/333 (49.5%) of respondents were aged over 65 yr. Almost two-thirds (198/316) had no formal educational qualifications and 166/332 (50%) were retired. The median disease duration was 13.3 yr [interquartile range (IQR) 6–25 yr]. The mean mHAQ score was 1.92 (S.D. 0.64). The mean fatigue VAS was 57.0 (S.D. 28.2) and mean pain VAS was 47.7 (S.D. 25.0). The mean RAI score was 21.24 (S.D. 5.9) and the mean psychological distress score was 2.0 (S.D. 0.69). Almost all (313/336 or 93%) of the respondents were taking DMARDs, and 189/313 (60%) reported having had an adverse reaction. The median number of DMARDs reported was 3 (IQR 2–5) over a median of 10 yr (IQR 4–19). The mean RA knowledge score was 51.8% (S.D. 23.3%).

**Beliefs about medicines scores**

The majority of the sample (74.3%) believed in the necessity of their medication for maintaining health (i.e. had scores greater than the scale midpoint; Table 1). Medication was considered important for both the maintenance of current health and for future health. However, overall 47.4% reported concerns about potential adverse consequences of taking their medication (i.e. scores greater than the scale midpoint; Table 1). Nearly 80% were concerned about potential long-term adverse effects of their medications. Another area of concern was becoming dependent upon medications (55.8% had scores greater than the scale midpoint). There was much less concern about the...
inconveniences and disruption to life’s routines associated with taking anti-rheumatic drugs or not having sufficient information about these.

The mean necessity score of 19.92 (S.D. 3.13) (Fig. 1) was significantly greater than the mean concerns score of 15.84 (S.D. 3.53; \( t = 16.59, P < 0.001 \)) (Fig. 2). The mean necessity–concern differential was +4.08 (S.D. 4.45). For 51 (15.6%) respondents their necessity score was lower than their concerns score (i.e. a negative necessity–concerns differential). For 23 (7.0%) respondents their necessity and concerns scores were equal.

**Associations of beliefs about medicines**

**Necessity of arthritis medication.** The mean necessity score for women (20.17, S.D. 3.22) was similar to that for men (19.43, S.D. 2.94; \( t = 1.98, P = 0.05 \)). Correlations between necessity scores and other variables are shown in Table 2. There was no association between the necessity score and the sociodemographic variables of age or level of formal education. Greater levels of pain, fatigue and physical disability (mHAQ) were associated with greater belief in the necessity of arthritis medications. Necessity scores also correlated positively with a sense of greater helplessness (RAI). Longer disease duration, longer DMARD treatment duration and greater numbers of DMARDs taken were all associated with greater belief in the necessity of medications, although the first two of these correlations were very weak. There was no correlation between knowledge about RA and necessity scores.

Once pain and fatigue were controlled for by partial correlation, there was no association between necessity scores and RAI (\( n = 268, r = 0.07, P = 0.23 \)). The association between mHAQ score and belief in the necessity of medication remained after controlling for pain and fatigue (\( n = 276, r = 0.24, P < 0.001 \)). Controlling for pain, fatigue and disability did not completely remove the association between necessity scores and number of DMARDs taken (\( n = 237, r = 0.17, P = 0.01 \)).

**Concerns about arthritis medication.** Concerns scores for men (mean 15.90, S.D. 4.00) and women (mean 15.88, S.D. 3.32) were similar (\( r = -0.03, P = 0.98 \)). Correlations of concerns scores with other variables are shown in Table 3. There was a very weak

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**Table 1. Percentage of respondents agreeing/strongly agreeing with Beliefs about Medicines Questionnaire statements**

<table>
<thead>
<tr>
<th>Percentage agreeing or strongly agreeing</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Necessity scale</strong></td>
</tr>
<tr>
<td>My health, at present, depends on my medicines (n = 337)</td>
</tr>
<tr>
<td>My life would be impossible without my medicines (n = 341)</td>
</tr>
<tr>
<td>Without my medicines I would become very ill (n = 341)</td>
</tr>
<tr>
<td>My health in the future will depend on my medicines (n = 342)</td>
</tr>
<tr>
<td>My medicines prevent me from becoming worse (n = 342)</td>
</tr>
<tr>
<td><strong>Concerns scale</strong></td>
</tr>
<tr>
<td>Having to take medicines worries me (n = 341)</td>
</tr>
<tr>
<td>I sometimes worry about the long-term effects of my medicines (n = 341)</td>
</tr>
<tr>
<td>My medicines are a mystery to me (n = 337)</td>
</tr>
<tr>
<td>My medicines disrupt my life (n = 337)</td>
</tr>
<tr>
<td>I sometimes worry about becoming too dependent on my medicines (n = 342)</td>
</tr>
</tbody>
</table>

\( n \) values indicate number of respondents answering each question.

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**Table 2. Associations of study variables with Beliefs about Medicines Specific Necessity scores**

<table>
<thead>
<tr>
<th>Variable</th>
<th>( n )</th>
<th>Pearson’s ( r^a )</th>
<th>( P )</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>325</td>
<td>-0.07</td>
<td>0.24</td>
</tr>
<tr>
<td>Formal qualifications</td>
<td>309</td>
<td>-0.08</td>
<td>0.19</td>
</tr>
<tr>
<td>Pain VAS</td>
<td>283</td>
<td>0.23</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Fatigue VAS</td>
<td>282</td>
<td>0.30</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Modified HAQ</td>
<td>334</td>
<td>0.36</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>RAI</td>
<td>322</td>
<td>0.23</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>RAI :</td>
<td>327</td>
<td>0.32</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Disease duration</td>
<td>321</td>
<td>0.20</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Number of DMARDs taken</td>
<td>279</td>
<td>0.26</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>DMARD treatment duration</td>
<td>290</td>
<td>0.15</td>
<td>0.01</td>
</tr>
<tr>
<td>RA knowledge</td>
<td>330</td>
<td>0.02</td>
<td>0.66</td>
</tr>
</tbody>
</table>

\( \text{RAI, Rheumatology Attitudes Index; RAI : medication scores omitted.} \)

\( a \) Zero order correlations.

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**Fig. 1. Frequency distribution for Beliefs about Medicines Questionnaire Specific Necessity Scale.**

**Fig. 2. Frequency distribution for Beliefs about Medicines Questionnaire Specific Concerns Scale.**
negative association with age, older people tending to report less concern. There was also a very weak negative correlation between level of formal education and concerns, more highly qualified people having lower concerns scores. Higher pain and fatigue scores were associated with greater levels of concern about taking medications. There was a moderate positive correlation between mHAQ score and concerns score. The correlation between concerns scores and RAI was strong, even when the three medications-related components of this index were omitted.

Although neither disease duration nor duration of DMARD treatment correlated with concerns scores, the greater the number of DMARDs taken the higher the concerns score. Those who reported having had adverse effects from treatment had higher concerns scores (mean 16.49, s.d. 3.68) than those who did not (mean 14.64, s.d. 2.98; \( t = 4.60, P < 0.001 \)). There was no association between knowledge about RA and concerns scores. The association between concerns scores and RAI remained after controlling for age, formal education, pain, fatigue and mHAQ score. The association between concerns score and mHAQ score was removed when pain and fatigue were controlled for (\( r = 0.002, n = 272, P = 0.975 \)). However, the correlation between concerns score and number of DMARDs taken remained after adjusting for pain, fatigue and mHAQ score (\( r = 0.189, n = 235, P = 0.003 \)).

The necessity–concerns differential. Factors significantly associated with a positive necessity–concerns differential were a lower RAI score (i.e. less helplessness) (\( n = 314, r = -0.28; P < 0.001 \)), less psychological distress (\( n = 325, r = -0.19; P < 0.001 \)) and better adherence (\( n = 322, r = -0.17; P = 0.002 \)).

### Relationship of drug adherence to beliefs about medications and to disease knowledge

Necessity scores for the adherent (\( n = 304, \text{mean 19.73, s.d. 2.78} \)) and non-adherent (\( n = 27, \text{mean 19.92, s.d. 3.18} \)) drug therapy groups were not significantly different (\( t = 0.291, P = 0.77 \)). However, concerns scores of the adherent group (mean 15.64, s.d. 3.51) were significantly lower than those of the non-adherent group (mean 17.88, s.d. 3.29; \( t = -3.136, P = 0.002 \)). The necessity–concerns differential was also significantly higher for the adherent group (mean +4.28, s.d. 4.5) than for the non-adherent group (mean +1.84, s.d. 3.57; \( t = 2.68, P = 0.008 \)). Knowledge scores for those who reported not taking their medications as directed (mean 45.12, s.d. 22.90) did not differ significantly from the scores for respondents who adhered to treatment (mean 52.51, s.d. 23.35; \( t = -1.58, P = 0.12 \)).

### Discussion

The main finding of our study was that three-quarters of people with RA have positive beliefs about the necessity of their medication. However, strong concerns about potential adverse effects, particularly long-term effects, were also expressed by almost half the respondents. In comparison with other chronic illness groups, the average necessity score was similar to that observed among asthmatic, renal dialysis and oncology patients ([2]. The average concerns score was similar to that of asthmatic patients, and notably higher than that for renal dialysis, oncology and cardiac patients ([2].

Two previous studies of medication beliefs of patients with rheumatological conditions also found that participants expressed strong concerns about potential adverse consequences of treatment ([6, 7]. In addition, the psychological impact of being withdrawn from a DMARD was considerable ([7]. We found that the greater the number of DMARDs patients had taken, the greater their concerns about medications, even after adjusting for pain, fatigue and disability. However, belief in the necessity of arthritis medication also positively correlated with the number of DMARDs taken and this association also remained after adjustment. It seems that with increasing numbers of DMARDs taken, patients' increasing concerns are counterbalanced by a greater perceived necessity. To our knowledge, only one previous study has used the BMQ to assess medication beliefs among patients with RA. Skingle investigated whether current beliefs were associated with past decisions to reject starting DMARDs ([16]. The group who had rejected DMARDs had significantly greater concerns scores, but necessity scores did not differ from those who had started treatment.

This is the first study of people with RA that examines potential associations of medication beliefs assessed using the BMQ. It is striking that sociodemographic factors (age, gender and formal education) showed no association with medication beliefs. In addition, there was no association between concerns scores and duration of disease or duration of DMARD therapy. There were weak correlations between necessity scores and disease duration and DMARD therapy duration, which were removed after adjusting for pain, fatigue and mHAQ score. These findings are perhaps not surprising in the light of a previous study of patients with RA which showed that psychological well-being and disease acceptance were independent of disease duration ([17].

We found that pain and fatigue were both associated with greater concerns scores and greater necessity scores. Physical disability (mHAQ) was associated with greater beliefs in the necessity of medications, independent of pain and fatigue. Although disability correlated with concerns about medications, this association was removed after adjustment for pain and fatigue. A previous study found no associations between willingness to accept treatment risk and disability or pain ([18].

The RAI measures perceived helplessness, and has been found to correlate with age, low education, HAQ and pain ([11]. RAI scores strongly correlated with concerns scores, even after controlling for age, education, pain, fatigue and disability. As this is a cross-sectional study, it is unclear whether helplessness increases concerns or vice versa.

There was no association between participants' knowledge about RA and their beliefs about arthritis medications. This is surprising as it could be expected that greater knowledge would be associated with greater beliefs in the necessity of medications and reduced levels of concerns about medications. A previous study found there was no difference in knowledge of steroid side-effects between those willing and those not willing to take steroids ([19]. One could infer that education does not appear to convince patients of the need for medications or reassure them about potential adverse effects. Lay beliefs are based not only on medical knowledge but also upon commonsense beliefs derived from other sources, and are also influenced by the interpretation.
of the impact of disease upon an individual [20]. Providing education about drug therapy alone is insufficient. The BMQ has the potential to be a useful clinical tool, as well as a research tool, in providing a focus for patients to discuss their beliefs. Specific concerns about long-term effects of RA medication and dependency were common, and thus need to be a focus of drug counselling.

Over half of our sample reported perceived adverse effects from medications, in line with rates reported in previous studies of patients with RA [6] and ankylosing spondylitis [21]. We found that those who had experienced DMARD adverse effects were more concerned about their medications. However, in this cross-sectional study we cannot conclude that adverse effects are responsible for medications concerns. In fact, longitudinal observations have shown that changes in RA symptoms and the incidence of medication side-effects are predicted by baseline somatization [22].

Complex psychological factors underpin adherence [4, 23]. Whether or not a patient will adhere to treatment appears to partly depend on their weighing up the perceived risks and benefits of treatment. An American study found that people with RA were generally unwilling to accept treatment risks [24]. Considering the necessity–concerns differential, a cost–benefit assessment, nearly a quarter of our respondents could be considered at risk of drug treatment non-adherence, their concerns about medications being as great as or greater than their beliefs in the necessity of their medications. This sizeable proportion is consistent with previous work showing levels of non-adherence to be 30–50% [4]. The BMQ has the potential to be clinically useful as a screening tool for the identification of non-adherence.

We also examined whether adherence was associated with peoples’ level of knowledge about RA. Knowledge scores for those who reported not taking their medications as directed did not differ from the scores for respondents who were adherent to treatment. We assessed factual and rather abstract knowledge. However, knowing facts about treatment does not necessarily result in changes in health behaviours, whereas self-management education, based on self-efficacy theory, has been shown to improve adherence with medication [25] and a self-efficacy scale [26] could be a useful additional item in future studies investigating medication beliefs.

Participants who reported not taking their medications as directed reported greater medication concerns than did those who were adherent, an association found previously [2, 27]. Interestingly, reported beliefs in the necessity of medications did not differ for the two groups. This indicates that we should perhaps concentrate more on reassuring patients about the safety of the treatments we prescribe, specifically reducing their fears about long-term effects of taking medications and dependency, than on convincing them of the benefits of treatment. Provision of standard drug information sheets to all patients with RA taking DMARDs is now widespread [28], and these of necessity focus on potential side-effects. It is up to the clinician to discuss these with patients in a way that is meaningful to them as individuals, otherwise there is a risk that this information may contribute to peoples’ concerns.

There are a number of limitations to this study. Firstly, its cross-sectional design prevents conclusions about the causality of the associations we observed. For example, we cannot be sure whether concerns about medications lead to non-adherence or whether non-adherence leads to greater concerns. Medication beliefs are dynamic [7, 29], an aspect that we could not address. Quantitative analyses such as this cannot capture all the facets that would be identified in a qualitative study, but the relatively large sample size gives us some confidence in the significance of our findings. However, our response rate was moderate, and it is likely that the medications beliefs of non-responders are different from those of the participants. Future studies should focus on this group.

Despite these caveats, it is clear that patients with RA express strong concerns about taking medications, particularly about long-term effects and dependency. The BMQ has the potential to be a useful tool in providing a focus for patients to discuss their medication beliefs. It could also be a useful screening tool in the identification of poor adherence, identifying concerns that can form the basis of counselling.

<table>
<thead>
<tr>
<th>Key messages</th>
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<tbody>
<tr>
<td>- Most people with RA have positive beliefs about the necessity of their medication.</td>
</tr>
<tr>
<td>- However, levels of concern are high and associate with helplessness and non-adherence.</td>
</tr>
</tbody>
</table>

The authors have declared no conflicts of interest.

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Clinical Vignette

An unusual case of extra bone formation

The patient, a 40-yr-old female with fibrodysplasia ossificans progressiva, first presented in her teens with pain and stiffness of her arms. Her symptoms progressed to involve her legs and axial skeleton and she became wheelchair-bound at the age of 40. Her condition continues to deteriorate.

The clinical features are of progressive ossification, which starts distally and affects axial muscles, ligaments, fascia, tendons and joint capsules (Fig. 1). Symmetrical digital abnormalities are present at birth and normally precede the soft-tissue ossification. The most common abnormality is hallux valgus with microdactyly of the first toes, with hypoplasia or synostosis of the phalanges. In some cases similar abnormalities are found in the fingers (Fig. 2).

A variety of treatments, including corticosteroid and bisphosphonates, have been used but none has been shown to slow disease progression.

It is a rare autosomal dominant condition (1 per 2 million) with variable phenotypic expression but complete penetrance. The gene responsible is found on chromosome 4q27-31 but no mutation has been identified. Although the exact mechanism remains unclear, there is overexpression of bone morphogenetic protein type 4 in circulating lymphocytes and in all cell types in the affected areas.

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Fig. 1.

Fig. 2.