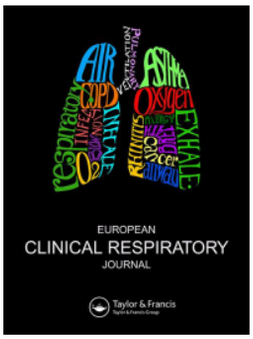

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The diagnostic trajectories of Danish patients with autoimmune rheumatologic disease associated interstitial lung disease: an interview-based study

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ABSTRACT

Background: Autoimmune rheumatologic disease associated interstitial lung diseases (ARD-ILD) are rare conditions and the association between ARDs and respiratory symptoms often goes unrecognised by ARD patients and general practitioners (GPs). The diagnostic trajectory from the first respiratory symptoms to an ARD-ILD diagnosis is often delayed and may increase the burden of symptoms and allow further disease progression.

The aim of this study was to 1) characterise the diagnostic trajectories of ARD-ILD patients and to 2) identify barriers for obtaining a timely ILD diagnosis based on the experiences and perceptions of both patients and healthcare professionals.

Method: Semi-structured qualitative interviews were conducted with Danish ARD-ILD patients, rheumatologists, pulmonologists and ILD nurses.

Results: Sixteen patients, six rheumatologists, three ILD nurses and three pulmonologists participated. Five characteristics of diagnostic trajectories were identified in the patient interviews: 1) early referral to lung specialists; 2) early delay; 3) delay or shortcut depending on specific circumstances; 4) parallel diagnostic trajectories connected late in the process; 5) early identification of lung involvement without proper interpretation. With the exception of early referral to lung specialists, all of the diagnostic trajectory characteristics identified led to delayed diagnosis. Delayed diagnostic trajectories resulted in patients experiencing increased uncertainty. Inconsistent disease terminology, insufficient knowledge and lack of awareness of ARD-ILD among central healthcare professionals and delayed referral to ILD specialists were main contributors to the diagnostic delay identified by the informants.

Conclusion: Five characteristics of the diagnostic trajectories were identified, four of which led to diagnostic delay of ARD-ILD. Improved diagnostic trajectories can shorten the diagnostic trajectory and increase early access to appropriate specialist medical care. Improved awareness and expertise in ARD-ILD across different medical specialties, especially among GPs, may contribute to more efficient and timely diagnostic trajectories and improved patient experiences.

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Introduction

Patients with autoimmune rheumatologic diseases (ARD) such as rheumatoid arthritis (RA), inflammatory myopathies and systemic sclerosis (SSc) frequently manifest with multiorgan involvement. The lungs are often involved in the form of interstitial lung disease (ILD) [1–4]. Autoimmune rheumatologic disease associated interstitial lung diseases (ARD-ILD) are often also known as connective-tissue-disease ILDs (CTD-ILD) but this term excludes patients with rheumatoid arthritis.

Several subtypes and phenotypes of ILD exist [5]. Some ILDs are characterised by a progressive pulmonary fibrosis (PPF) phenotype and are associated with impaired quality of life and survival [6,7], while other types have less impact on morbidity and mortality. However, early diagnosis is equally important across all the different subtypes of ILD, in terms of avoiding exposure and initiating treatment. Early immunomodulatory treatment has the potential to improve patients with a more inflammatory phenotype [8,9] or to reduce disease progression, whereas antifibrotic therapy can slow down irreversible changes in patients with PPF [10–13].

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Current research in non-ARD ILDs has shown that significant diagnostic delay is often caused by factors such as misdiagnosis, underreporting of ILD features on imaging and prolonged referral time to ILD specialists, even when ILD is reported [14–17]. Similarly, previous studies on ARD-ILD have found that the time from symptom debut to diagnosis is delayed and that diagnostic delay is associated with disease progression and increased mortality [18,19].

Only a few studies have examined how patients and healthcare professionals (HCPs) experience and perceive diagnostic trajectories and the barriers related to the pre-diagnostic period in patients with ARD-ILD. Previous research has primarily focused on SSc-ILD [20,21,22].

The aim of this qualitative study was to 1) characterise the diagnostic trajectories of ARD-ILD patients based on their experience and to 2) identify barriers for obtaining a timely ILD diagnosis based on the experiences and perceptions of both patients and healthcare professionals.

Data and methods

Design

Our study had a qualitative design using semi-structured interviews. This method provides insight into the experiences of the patients from the time they experienced the first symptoms to the time they received the ARD-ILD-diagnosis. It also allows for a focused exploration of the main barriers in the diagnostic trajectories from the perspective of patients and healthcare professionals.

Our study was based on a project description outlining the phases and elements of the study as is customary for qualitative studies.

Recruitment procedure

The primary inclusion criterion for patients was a diagnosis of ARD-ILD in 2019, 2020 or January-May 2021. A pulmonologist from each ILD centre invited patients with ARD-ILD to participate in the study. Interested patients gave written consent to be contacted by telephone by the first (M. B. Johansen) or last author (H. M. Martin) in order to confirm their interview participation and make an interview appointment. The interviewers had no access to patient records.

Health care professionals (HCPs) with different levels of experience and geographical locations were selected based on their experiences with ARD-ILD patients, diagnostic trajectories and potential barriers. ILD specialists were recruited based on their experience in the field and were all working at a tertiary ILD centre. Rheumatologists from tertiary and secondary hospitals were recruited through either open invitations to the department via email or via invitations sent to specific persons based on suggestions from the participating ILD specialists.

All informants received written information about the study and the data protection procedures followed by VIVE – The Danish Center for Social Science Research. VIVE is an independent research and analysis centre operating under the Danish Ministry of the Interior and Housing and conducts research and analysis projects in all the major aspects of welfare and the Danish welfare state. The interview data was stored in accordance with the general data protection regulations (GDPR). All interviews were performed by M. B. Johansen and H. M. Martin.

The interviews

A semi-structured interview guide that addressed aspects of the diagnostic trajectory was developed based on inspiration from a Danish study on the diagnostic trajectories of IPF patients [23] and expert input from an advisory board of the study consisting of representatives of the patient associations (Danish Rheumatology Association, Danish Lung Association and Danish Association for Systemic Sclerosis), medical experts in the field of ILD and rheumatology and an experienced qualitative researcher with knowledge in the area of diagnostic delay.

Semi-structured interviews are particularly relevant in qualitative studies that have a narrow research focus. The researcher can ask questions relating to the specific topic, while inviting the informant to contribute information that were unforeseen in the interview guide. Semi-structured interviews will cover the central topics and allow for emergent themes [24]. The semi-structured interview format also left room for the interviewer to ask informants for more details and to elaborate on any particularly interesting themes.

The interviews were conducted from February to May, 2021. M. B. Johansen conducted all nine HCP interviews and nine patient interviews, while H. M. Martin conducted seven patient interviews. The ILD specialists and ILD nurses were interviewed in pairs. Owing to the COVID-19 situation, the interviews were performed in a virtual meeting format (five HCP interviews) or as telephone interviews (16 patient interviews and four HCP interviews) according to informant preference.

The interviews were audio-recorded and detailed summaries were written by the interviewer immediately after the interview using the extensive notes taken during the interview. The recordings were visited for specific details and exact wording for use in citations.

The interviews were initially thematically coded [25,26] and similarities and differences in the patients' experiences in their diagnostic trajectories were identified. This led to the identification of central routes and points of passage of the diagnostic trajectories. Furthermore, it enabled the identification of the characteristics connected to diagnostic delay presented in the results section. The analysis was supported by Nvivo software. NVivo is a qualitative data analysis (QDA) software which enables the organization and analysis of qualitative data. Through categorization and comparison, the researcher can examine connections and relationships within and between the transcribed interviews.

Ethics

The study complied with ethical principles for medical research as described in the Helsinki Declaration. The study was registered in the VIVE internal GDPR register in accordance with standard procedures. The patients were informed of the study verbally and in writing and oral consent to participate was obtained. The informants gave permission to audio-record the interviews. They were guaranteed anonymity in the final report. All rules were followed in order to safeguard personal data. According to Danish legislation, ethics committee approval is not required under these circumstances.

Results

Informant characteristics

Sixteen patients were recruited from the three tertiary ILD centres in Denmark. The participating patients represented different ages, genders and ARD-diagnoses, as shown in Table 1. None of the participating patients had any known cognitive deficits.

The patient interviews lasted between 16 and 78 minutes. The wide range of durations reflects the complexity of patient trajectories. For instance, the two very short interviews (<20 minutes) in the data material were done with SSc-ILD patients who had experienced fast and uncomplicated diagnostic trajectories.

Twelve HCPs with different levels of experience and geographical locations were included. Three pulmonologists ILD specialists, three ILD nurses and six rheumatologists participated.

Impact of the COVID-19 pandemic

The majority of the patients interviewed (11 of 16) were diagnosed with ARD-ILD during the COVID-19 pandemic, but their initial contact to the healthcare system took place before the COVID-19 pandemic. The ILD specialists from the tertiary ILD centres reported that fewer pulmonary function tests were performed by GPs during this time, and that there was prolonged waiting time in hospitals as a result of outpatient clinics and ILD centres being partly closed-down during the pandemic. However, they also stated that referrals suspected for ILD were prioritized corresponding to national guidelines for assessment. The ongoing pandemic, the changed deployment of personnel, and accumulation of referrals may have slightly affected the waiting time for assessment. However, the patients had the impression that COVID-19 had not had a major influence on their diagnostic trajectories. Although the pandemic may have increased the length of the diagnostic delay for some of the patients it has not affect the diagnostic trajectories or barriers involved.

Characteristics of the diagnostic trajectories

The characteristics of the patients' diagnostic trajectories from first respiratory symptoms to obtaining an ARD-ILD diagnosis were analysed. The description of disease trajectories is primarily based on patient experience. However, these descriptions aligned with the core concerns that HCPs identified in the interviews.

Patient trajectories were very heterogeneous. Table 2 shows the sequence of diagnoses and the estimated

Table 1. Patient characteristics.

Gender	Age	ARD
Females: 12	41–50: 2	Systemic sclerosis: 8
Males: 4	51–60: 5	Rheumatoid arthritis: 5
	61–70: 4	Myositis: 3
	71–80: 3	
	81–90: 2	

Table 2. Patient trajectories.

Sequence of diagnoses	Patients (No.)	Time from first respiratory symptoms to ILD diagnosis (months)	Patients (No.)
ARD prior to ILD	4	0–6	5
ILD prior to ARD	3	7–12	4
ARD and ILD in close proximity	9	13–18	2
		18+	5

time from the patient experiencing their first respiratory symptoms to the ILD diagnosis. Nine of the 16 patients were diagnosed with ARD and ILD in close proximity to each other, whereas seven of the patients either had their ILD or ARD diagnosis first. Seven of the 16 patients had diagnostic trajectories lasting more than one year, with four years being the longest diagnostic trajectory. Five diagnostic trajectories were identified based on the patient interviews and illustrate the many trajectories from first respiratory symptom to ARD-ILD diagnosis. We identified different characteristics that played a significant role in the overall sequence of the diagnostic trajectories. The characteristics were not mutually exclusive. As seen in Figure 1, the different trajectories involve HCPs from several medical specialties. The descriptions below contain direct quotes from the patient interviews, in order to illustrate the different trajectories.

First diagnostic trajectory: early referral to lung specialists

A few of the patients reported fast and effective diagnostic trajectories. These trajectories typically involved few medical specialties and minimal waiting time. A typical characteristic was early referral from a GP, the first point of contact due to respiratory symptoms for 12 of the 16 patients interviewed, to a department of respiratory medicine, where a lung specialist initiated the relevant examinations without significant waiting time. Cases of SSc-ILD were typically diagnosed following this type of diagnostic trajectory:

> I had been coughing for three months and that's why I went to see my GP. I had no idea what it might be but I talked to my GP who referred me to have a chest X-ray. It showed lung fibrosis and I was referred to a lung specialist at the hospital. The doctor asked many questions and I told him that I had had white

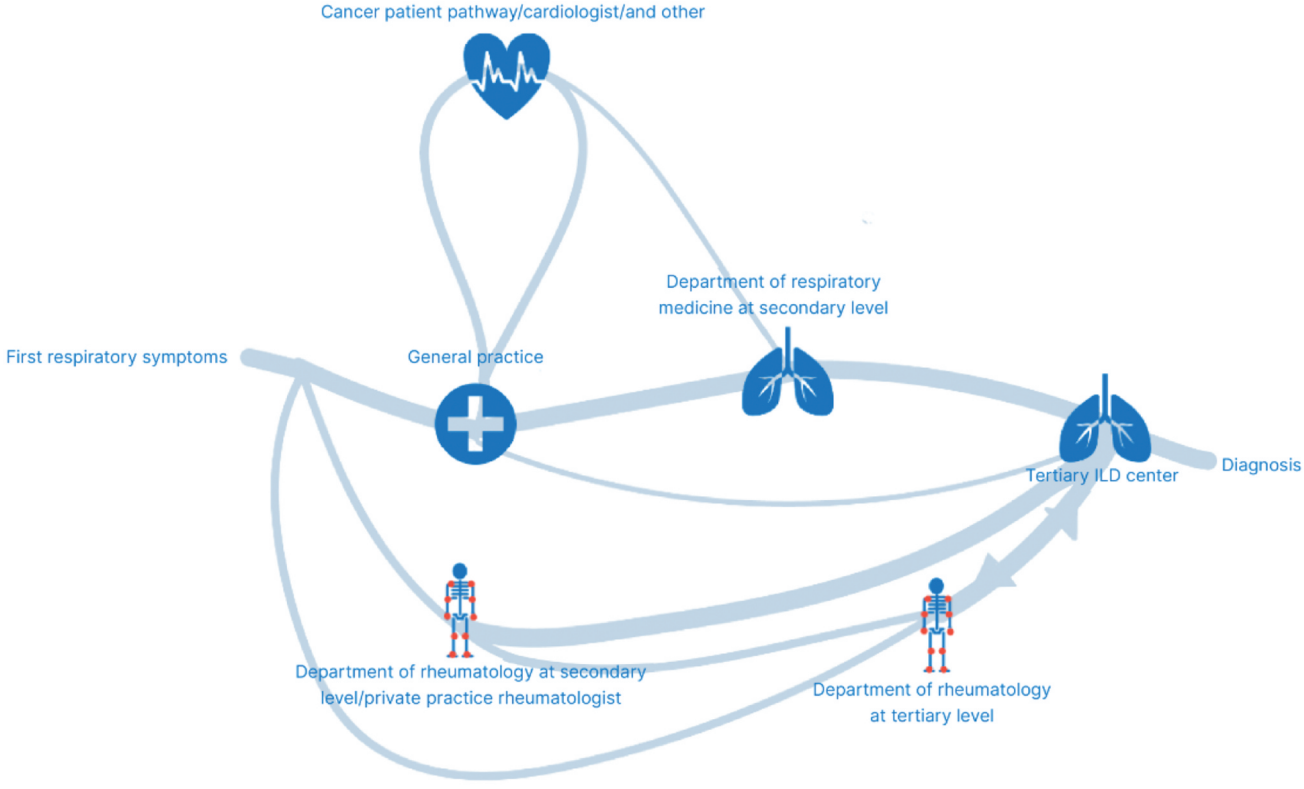

Figure 1. Map of diagnostic trajectories based on patient interviews. The thicker lines show the most common routes identified in the data set, while thin lines represent the less common routes (figure adapted from Johansen & Martin [27]).

> hands and feet for years. This caused him to suspect systemic sclerosis. They took some blood samples, which pointed at systemic sclerosis. The diagnosis was given after the blood samples has been analysed and doctors had examined my heart, gastrointestinal tract and lungs. They confirmed systemic sclerosis.

Only one of the SSc patients had a diagnosis of SSc prior to their ILD diagnosis and ILD and SSc were typically diagnosed concomitantly.

Other examples of early referral to pulmonologists were reported by two patients who contacted their rheumatologist due to respiratory symptoms, resulting in referral to a pulmonologist and an ILD diagnosis. Furthermore, there were two patients with no respiratory symptoms in this category, where thoracic imaging performed on other indications showed ILD.

Second diagnostic trajectory: early delay

A second characteristic identified was early delay in the diagnostic trajectory, defining the course and duration of the disease. According to the patients interviewed, the delay was caused, for instance, by GPs repeatedly prescribing therapy like antibiotics or medication for other respiratory conditions like asthma and COPD, even though the patients did not report any beneficial effects. Some patients found that their GPs appeared to be obstacles for further referral in cases of ongoing respiratory symptoms despite treatment attempts. However, the diagnostic process often accelerated when the GP eventually referred the patient to another specialist:

> I told my podiatrist – who is also a registered nurse – that it hurt when I breathe and that I had pain in my feet. They were very swollen and the skin on the feet had changed. My podiatrist looked at my feet and told me, "It looks like you have rheumatoid arthritis. You need to see a doctor!" I saw my GP but he just listened to my lungs and took some blood samples. He thought I had pneumonia and gave me a prescription of antibiotics […] he gave me antibiotics more than ten times […] Finally, I said, "Now it's been more than three years – you need to refer me to be seen by a specialist". Then he referred me to a rheumatologist […].

The early delay resulted in a prolonged diagnostic trajectory and the patients experienced a high degree of frustration, anxiety and self-doubt in relation to their symptoms as time elapsed, particularly if they also experienced worsening respiratory symptoms during the diagnostic delay.

Third diagnostic trajectory: delay or shortcut depending on circumstances

A third characteristic in the diagnostic trajectory was when the GPs referred patients for further investigation, based on the suspicion of other, non-ARD-ILD respiratory diseases. For example, referral for a computed tomography of the thorax (CT) on the suspicion of lung cancer or referral to a cardiologist for suspected cardiac disease. Such referrals could either result in delays or in a shortcut to an ILD diagnosis, depending on the specific circumstances, e.g. whether the radiologist identified and described the ILD findings on the chest CT and whether the cardiologist who ruled out a cardiac disease directly referred the patient to a pulmonologist or returned the patient to the GP. Sometimes, patients play a defining role, as in the following case where the patient was involved in creating a shortcut to an ILD diagnosis.

> I went to see my GP last spring. He suspected I might have cancer so I was sent for a diagnostic cancer examination. At the hospital, I was told that it was not cancer but there [at the specific hospital department], they only deal with cancer [so I was sent back to my GP]. In the test results, there was a note […]. I called my GP to ask what the letters meant. At first, he didn't think they meant anything but after a couple of days, he called me and said, "I've read about it. It's something to do with your lungs". He then referred me to a pulmonologist and I had a new lung scan done. Then, the doctor there referred me to yet another hospital because they are specialists there […].

Seemingly small details, such as when the patient notices things and enquires about the meaning of a test, can have substantial impact on the diagnostic trajectory.

Fourth diagnostic trajectory: parallel diagnostic trajectories connected late in the process

A fourth diagnostic trajectory characteristic was identified in interviews with patients diagnosed with both ARD and ILD in parallel trajectories, where both conditions were diagnosed separately and where a link between ARD and ILD was established late in the diagnostic trajectory.

> I've had joint pain for several years but all my blood tests came back clear, so I've gone years without a diagnosis. I've been to the rheumatology department many times but it has always been disregarded. […] I ended up getting very ill before they could see anything in my blood and when I eventually got a diagnosis, the rheumatoid arthritis had been able to develop heavily before I eventually began treatment […]. I'd just started treatment when I got a pneumonia and could hardly breathe […]. I was hospitalised and at the hospital, they did a CT scan, where they saw both inflammation and some shadows in the lungs […]. I was referred to pulmonologists at another hospital. There was a couple of months'

> waiting time to be seen there. They looked at the scan, did a spirometry and gave me an asthma spray for the dyspnoea, which they linked to the pneumonia. They gave me an appointment for another spirometry half a year later [...]. My rheumatologist then referred me to another hospital [more specialised] because he couldn't get my RA under control because I had to stop methotrexate because of the lung problems. So, he made sure that I was referred to the other hospital for both the lung problem and the RA. This was in late 2019 and I had an appointment in March 2020 [...]. There, I was told that the alterations in my lungs were RA-related.

Diagnostic trajectories with parallel diagnostic pathways at different departments typically resulted in diagnostic delay as several actors were involved with accumulated waiting time. At the same time, the patients experienced a growing uncertainty and often worsened symptom burden during the process.

Fifth diagnostic trajectory: early identification, late interpretation and follow-up

Finally, some diagnostic trajectories were initiated with sometimes random detection of preclinical ILD imaging findings that were not acted upon before the patients developed respiratory symptoms. The reasons given by the patients included that the pulmonary imaging findings were considered to be adverse events caused by therapy for their rheumatologic disease or that they did not know why the suspicion of ILD was dismissed by the doctors.

> It has been almost 30 years since my rheumatoid arthritis began. I started having pain in my fingers and toes but I quickly got treatment for it. One day [several years later] I was walking home with a friend. I was usually the one walking ahead but all of a sudden, I was short of breath and had difficulty breathing [...]. My GP told me she suspected that it was connected to the rheumatoid arthritis so she referred me for a chest x-ray of my lungs and I was told that something had changed in the lower part of my lungs. They thought it had to do with the methotrexate, so they stopped it. This was around 2005. But they didn't do anything about my lungs and didn't examine it further [...]. When I started on biological therapy, the lung symptoms improved and I felt better. Then, three years ago, I suddenly started to suffer from shortness of breath again [...]. My GP referred me to the pulmonologists at the hospital, where they thought I had asthma. However, I got so sick from the treatment that I had to be hospitalised. Afterwards, my GP again referred me to the pulmonologists at the hospital, who then [did a HRCT] found out that I had lung fibrosis.

In general, patients diagnosed with ARD before ILD were not aware of the possible association between the respiratory symptoms and their rheumatologic disorder. Therefore, patients did not associate the two diseases upon diagnosis of each. Typically, patients contact their GP rather than the rheumatologist with their respiratory symptoms, as they do not connect the two and this probably contributes to the diagnostic delay.

The characteristics presented in this section may coexist in a patient trajectory and thus create even more complex and protracted trajectories. In the text that follows, we will present three barriers that contribute to diagnostic delay. These are circumstantial elements that influence diagnostic trajectories more generally.

Barriers contributing to diagnostic delay

There are several contributing factors that can lead to different kinds of delay, as presented above. These factors are characteristic of the organizational and healthcare professional contexts in which the patient trajectories take place. Some of the factors are described in the patient interviews and confirmed in interviews with HCPs. Others are primarily described in HCP interviews and are more related to expert knowledge and experience of ARD-ILD, alongside multidisciplinary collaboration. Here we present three main barriers identified in both patient and HCP interviews.

Insufficient knowledge and awareness of ARD-ILD among HCPs

Across the interviews with HCPs, the main recurring theme was a lack of knowledge of the association between ARD and ILD among the HCPs typically involved in the diagnostic trajectories. While the association between ARD and ILD is well known among specialists in rheumatology and pulmonology at tertiary centres, interviews with patients reported that this is not always the case among GPs and HCPs at secondary centres. Furthermore, only one of the patients who received their ARD diagnosis before their ILD diagnosis was aware of the risk of lung involvement. Interviews with the HCPs indicate that this lack of knowledge is the main barrier contributing to the diagnostic delay experienced by many ARD-ILD patients.

Confusing and variable disease terminology and understandings

Another recurring theme from the patient interviews were difficulties related to disease terminology. Many patients reported that they did not know or understand

that they had lung fibrosis (as the ARD-ILD was often termed in the patient interviews) ahead of participating in this study. Across the patient interviews, the terminology used by the patients to talk about their ARD-ILD varied significantly. This reflects the variation in terminology used by the HCPs. The patients were fully aware of their respiratory symptoms but not all patients had linked their ARD to their respiratory symptoms. This was indicated by some of the terms used by the patients e.g. *scar tissue in the lungs, polymyositis in the lungs, arthritis-related changes in the lungs, fibrotic alteration of the lungs*. However, a few patients used the term *lung fibrosis*. Others, primarily patients with SSC, considered ARD and respiratory symptoms as fully integrated and did not make a distinction between the two.

Multidisciplinary collaboration

Multidisciplinary collaboration is another central theme in the interviews with the HCPs. The HCPs interviewed considered MDTs to play an important role in achieving timely diagnosis. In a specialist health care system, collaboration across disciplines and departments can be challenging e.g. the interviewees pointed to the issue, that HCPs involved in different phases of the diagnostic trajectory may not have full insight into decisions and activities initiated elsewhere. Moreover, they argued that the complexity of the diagnostic process demands precise and close communication and knowledge of the roles of different HCPs involved in the diagnostic trajectory. The HCPs reported that when these elements are not present, there is a risk of delay of the diagnostic process e.g. due to late referral to ILD specialists. Furthermore, both formal and informal multidisciplinary collaboration is dependent on mutual respect and trust in one another's' specialist knowledge in order to be productive.

Discussion

The current qualitative interview study sheds light on the heterogeneous and complex diagnostic trajectories that can result in significant diagnostic delay for patients with ARD-ILD.

For many patients, it took more than 12 months from their first respiratory symptom to a final diagnosis of ARD-ILD. Similarly, patients with various underlying rheumatic diseases interviewed in a multinational qualitative study reported a median diagnostic delay of 11 months [28]. We found two types of delay described by patients: 1) the accumulated delay across the diagnostic trajectories caused by inappropriate referrals to non-relevant specialists and accumulated waiting time from referral to specific tests or examinations e.g. imaging; and 2) early onset of delay caused by late GP referral to specialists or delay caused by capacity-related waiting times for specific tests or examinations. Patients typically experienced increased concern and anxiety during the waiting time, and this was further pronounced when they simultaneously experienced an increased burden of symptoms. Interestingly, referral to other specialists could impact the diagnosis in either a positive or negative direction, or in other words, could constitute a shortcut or another delay, depending on the level of ILD knowledge and awareness among these specialists.

Our study points to several barriers and potential routes for improving diagnostic trajectories. Previous surveys on educational aspects of rare lung diseases have similarly pointed out the main challenges for HCPs, i.e. lack of diagnostic guidelines, delay or inability to make a definite diagnosis, doubts about referral procedures, and lack of education on how to communicate information on diagnosis and treatment plans with their patients [29]. A central result of our study is the lack of HCP and patient knowledge and awareness of the association between ARD and lung involvement. This lack of knowledge and awareness underlies many of the delay-associated characteristics of the diagnostic trajectories identified in this study. Similar findings of patients often being unaware of the potential relationship between respiratory symptoms and ARD have previously been reported [28]. This study also reported that patient symptoms, lung function and quality of life worsened, adding to patients' experience of increased anxiety and distress.

Many patients report to be treated repeatedly for respiratory infections or more common lung diseases like COPD, heart failure and asthma even though they report no improvement, a tendency that previously has been observed also in patients with idiopathic pulmonary fibrosis (IPF) based on prescription data and patient interviews [15,30]. Similar reasons for delay were also reported in an IPF cohort, where many patients experienced multiple erroneous differential diagnoses like asthma, emphysema and COPD, resulting in a diagnostic delay of more than one year for the majority of patients before they were diagnosed with IPF [15,31]. ARD-ILD are rare diseases and less than 1,500 Danish patients from a background population of 5.83 million were diagnosed with ARD-ILD between 2000–2015 [32,33]. Therefore, most GPs will only see very few patients with ARD-ILD during their career. However, this emphasises the importance of performing pulmonary function tests to prove or rule out

obstructive lung diseases and to refer patients with respiratory symptoms for further diagnostic work-up by pulmonologists if investigations are incompatible with more common lung disease such as COPD or persistent 'pneumonia' without effect of antibiotic treatment.

Early diagnosis can also be improved by making radiologists more aware of the radiological signs of ILD, particularly avoiding general unspecific terms like 'chronic lung changes'. Multidisciplinary evaluation of CT scans with relevant information from the referring physician is key and likely to result in more timely diagnosis of ARD-ILD.

The lack of awareness and knowledge of the risk of lung involvement in patients with ARD emphasises the need for education for both patients, GPs, radiologists, as well as community rheumatologists and pulmonologists. Only a few of the patients were aware of the risk of lung involvement due to their ARD and some patients suggested that increased awareness among patients with ARD also may shorten the diagnostic trajectory. Thus, increased patient information or even patient schools teaching patients about the potential risk of lung and other organ involvement in ARD might facilitate improved collaboration and awareness for both patients and HCPs and thus increase the likelihood of timely and early ARD-ILD diagnosis.

The lack of consistent disease terminology identified in this study is another problem, as patients will meet several HCPs throughout their diagnostic trajectory. The HCPs may use different terminology which the patients may adopt and use themselves to understand and explain their disease experience. This may apply to that for some ILD subtypes, no real layman's term exist, and in such incorrect terms are often used, e.g. 'lung fibrosis' for ILD subtypes dominated by interstitial inflammation in which lung fibrosis does not exist. The patients expressed that the use of different terminology contributed to confusion, concern and anxiety in an already complex diagnostic trajectory. Moreover, the inconsistent terminology might also indicate an increased risk of unclear communication and misunderstandings between patients and HCPs. A commonly accepted nomenclature for ARD-ILD and more standardised communication may minimise patient concern and uncertainty and simultaneously improve awareness and timely referrals among HCPs. Moreover, a common understanding and translation of ARD-ILD subtypes may increase the patients understanding of their diseases.

The results of this study also emphasize the need for improved diagnostic tools and collaboration across the disciplines involved in the diagnostic trajectory. Evidence-based guidelines for diagnosing and treatment of all ARD-ILDs have not yet been developed. However, algorithms for the diagnosis and treatment of SSc-ILD have been developed, and baseline HRCT and pulmonary function test at SSc diagnosis is now recommended as part of the diagnostic criteria for SSc [34]. Though formalised systematic awareness in the format of a screening programme exists for SSc-ILD, recommendations for screening programmes are lacking for patients with other ARDs. Proposals for screening algorithms for RA-ILD has recently been published [35,36]. The specialists who were interviewed had different perceptions of the benefit of screening in RA based on the large number of patients, the low prevalence of RA-ILD and concerns of burdening patients with unduly worries. In contrast, other HCPs found it highly relevant to identify ARD-ILD as early as possible to allow for early intervention. There was no consensus on the choice of screening, with the exception of systematic focus on respiratory symptoms and smoking cessation.

The HCPs interviewed stressed multidisciplinary collaboration between rheumatologists and pulmonologists as a core element with the potential to improve the diagnostic trajectories of ARD-ILD patients. The HCPs described the potential of both formal and informal multidisciplinary collaborations. Furthermore, they emphasised the benefits of multidisciplinary collaboration, both at the same level of specialisation and across specialisation levels. Early referral of patients suspected to have ARD-ILD to highly specialised ILD-centres was regarded as one of the most important elements for a timely diagnosis.

Conclusion

Our study showed that diagnostic trajectories from the first respiratory symptom to ARD-ILD diagnosis are characterised by heterogeneity and often result in diagnostic delay. Five characteristics of the diagnostic trajectories were identified, four of which led to diagnostic delay of ARD-ILD. Multiple diagnostic barriers are embedded in the organizational and professional context in which ARD-ILD trajectories take place. Some of the key actions for reducing diagnostic delay should involve improved education among the various actors involved. A common terminology among patients and HCPs and clearer information and communication with patients and caregivers is essential. Improved diagnostic trajectories will shorten waiting time and increase early access to appropriate specialist care. Improved awareness and expertise in ARD-ILD across different medical specialties,

especially among GPs, may contribute to more efficient and timely diagnostic trajectories and improved patient experiences.

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