SCLERODERMA LUNG DISEASE: WHAT THE PATIENT SHOULD KNOW

Lung disease can be a serious complication of scleroderma. The two most common types of lung disease in patients with scleroderma are interstitial lung disease, also called pulmonary fibrosis, and pulmonary hypertension. These pulmonary complications can occur in patients with limited or diffuse scleroderma. Although these are the most common manifestations, other less common forms of pulmonary disease can also occur in scleroderma (see the table below).

Types of Scleroderma Lung Disease

- Interstitial Lung Disease
- Pulmonary Hypertension
- Airways obstruction
- Aspiration
- Extrinsic restriction
- Lung cancer
- Muscle weakness
- Pleural Effusions
- Pneumonia
- Pneumothorax
- Pulmonary Hemorrhage
- Sarcoidosis

ASSESSMENT OF SCLERODERMA LUNG DISEASE

It is important to note that patients may have significant pulmonary involvement from their scleroderma before signs and symptoms appear. **Therefore, it is important to have routine screening for possible pulmonary involvement, in particular interstitial lung disease and pulmonary hypertension.**

Symptoms

- Chest pain, particularly on exertion, is a potentially serious symptom and should be evaluated promptly by your physician. Chest pain may be a warning sign that you are at risk of having a heart attack. Chest pain can also occur in patients that have pulmonary hypertension, or it may be a symptom of gastroesophageal reflux (GERD).

- Cough can be seen in patients with interstitial lung disease, pulmonary hypertension or with airways disease. However, the most common cause of chronic cough is gastroesophageal reflux disease, which is very common in patients with scleroderma. It is important to note that many patients with GERD-related cough do not have typical symptoms of GERD such as indigestion or heartburn. Typically, in scleroderma patients the cough is non-productive.

· Light headedness or fainting *(syncope)* with exertion is also a very serious symptom that should prompt immediate evaluation by your physician. It may be a result of pulmonary hypertension or a consequence of too little oxygen secondary to severe interstitial lung disease.

· Muscle weakness can be seen in patients with scleroderma, particularly in those patients who have an overlap with polymyositis or dermatomyositis. Patients with muscle weakness of their arms and legs can also have muscle weakness of the respiratory muscles.

· Palpitations (heart racing or fluttering) may be a symptom of pulmonary hypertension or heart disease and should be evaluated promptly by your physician.

· Shortness of breath, also called *dyspnea*, is the most common symptom in patients with scleroderma lung disease and should be promptly evaluated by your physician. However, patients may have significant pulmonary involvement without this symptom. Often patients gradually decrease their activity to avoid the unpleasant sensation of shortness of breath without even realizing it. ***It is important to screen for scleroderma lung disease even if you do not have shortness of breath.***

· Swelling is also referred to as *edema*. Lower extremity swelling is a potentially serious symptom and should be evaluated promptly by your physician. Swelling can occur in patients with pulmonary hypertension, very advanced interstitial lung disease and in patients with left-sided heart disease (congestive heart failure). Some medications may also cause leg edema.

· Wheezing is not common in scleroderma, but can be seen in patients with scleroderma-related airways disease, or in patients with a history of asthma or smoking. Airways disease is more common in scleroderma patients who also have features of rheumatoid arthritis or Sjogren's syndrome. Wheezing can also be a symptom of gastroesophageal reflux disease.

Tests Your Doctor Might Order

Pulmonary Function Tests

Pulmonary function tests (PFTs) are performed by blowing in a tube that is connected to a computerized machine.
It is important that you form a tight seal with the mouthpiece. Often patients with scleroderma need to use a mouthpiece made for children because they cannot open their mouth very wide.

Numerous measurements can be made during pulmonary function testing and can be suggestive of a specific diagnosis.

6 minute walk testing (6MWT) is simple and reproducible and correlates with findings on more formal exercise testing. It has been also shown to correlate with hemodynamics measured during heart catheterization, functional classification, prognosis and even survival. The 6MWT is routinely used in studies of pulmonary hypertension. During this test, you are asked to walk as far as you can during six minutes. You are allowed to rest if you need to. While doing this test, your heart rate, blood pressure, oxygen level, degree of shortness of breath and other symptoms are monitored. Ask your doctor to use a forehead probe to monitor your oxygen level as it is frequently difficult to detect oxygen levels with a finger probe in patients with Raynaud phenomenon.

Table: Recommendations for Pulmonary Function and Exercise Testing
There are no official recommendations for pulmonary function and exercise testing in patients with scleroderma. However, many experts recommend the following:

- **Baseline** complete set of pulmonary function tests (spirometry, lung volumes, diffusion)
- **£ 5 years of Scleroderma**
  - PFTs and 6MWT every 3-6 months
- **> 5 years of Scleroderma**
  - PFTs and 6MWT every 6-12 months

Chest radiography (Chest x-ray) is not a sensitive test for scleroderma lung disease. This means that you could have significant lung disease and still have a normal chest x-ray. However, there are features on the chest x-ray that should prompt your physician to do further testing

Chest Computed Tomography Scan (CT) is a special type of x-ray that allows one to see more details of the lungs. During a CT, you lie on your back and slide through a machine that looks like a giant doughnut. You may also be asked to also lie on your stomach (prone). Images are typically taken while holding your breath in inspiration. However, images may also be taken during expiration to see if air is trapped in your lungs. Finally, you may be given contrast dye in order to get a better look at the blood vessels in your lungs and to look for evidence of blood clots. A high resolution chest CT (HRCT) (or thin-slice CT) is considered the gold standard for evaluation for interstitial lung disease. Findings that are suggestive of interstitial lung disease include ground glass opacities (also called "alveolitis"), which looks like a haziness over the lung; septal thickening, which are extra lines in the lung because of thickening of the interstitium**;** and honeycombing, which represents holes in the lung. Ground glass opacities are suggestive of inflammation in the lungs that may be reversible, whereas honeycombing usually represents irreversible fibrosis.

Bronchoalveolar lavage (BAL) by bronchoscopy is sometimes done to look for inflammation or evidence of infection in the lung. A fiberoptic scope is passed through your nose or mouth down into your airways. Small amounts of sterile saline are then injected in an area of your lung and suctioned back out and sent for laboratory analysis**.** Although bronchoscopy and BAL to look for infection is a routine procedure, the analysis of the fluid for the presence of inflammatory cells (alveolitis) needs to be done by experienced personnel, usually at a scleroderma or pulmonary referral center.

There are no official recommendations for echocardiogram testing in patients with scleroderma. However, many experts recommend the following:

- Baseline and yearly screening echocardiogram
- Patients with pulmonary hypertension
  - Echocardiogram every 3-6 months
  - Echocardiogram if deterioration in clinical status

Echocardiogram is actually an ultrasound of your heart. By rubbing a probe across your chest, the cardiologist and can look to see how well your heart is pumping, see if your heart valves are working, evaluate the size of the various chambers of the heart, see if there is fluid around the heart (pericardial effusion) and estimate your pulmonary artery pressure.

Echocardiogram

Heart catheterization

- **Right heart catheterization** is essential for the evaluation of pulmonary hypertension. The cardiologist will thread a wire through either your femoral vein in your groin or internal jugular vein in your neck to the right side of your heart and into your pulmonary artery in order to measure the pressure on the right side of your heart and in your lungs. The right heart catheter can also be used to measure your cardiac output (how well your heart is pumping) and can estimate the pressures on the left side of your heart. ***A right heart catheterization needs to be done by an experienced cardiologist or pulmonologist* before *starting medicine for pulmonary hypertension.***

- **Left heart catheterization** is usually performed by threading a wire through your femoral artery in your groin. The cardiologist will do a left heart catheterization if he/she is looking for blockages of your coronary arteries, or needs to measure pressures on the left side of the heart in conjunction with pressures on the right side of the heart. A left sided heart catheterization is also sometimes needed in patients with heart valve problems.

Ventilation/Perfusion Scan (V/Q scan) is a special nuclear x-ray to see if areas of your lung that get air also are getting blood. A mismatch or defect suggests a blood clot. This test should be considered in patients with pulmonary hypertension because if blood clots in the lungs are identified, they can sometimes be surgically removed and the pulmonary hypertension cured.

Lung Biopsy is rarely indicated in patients with scleroderma and is especially risky if you have pulmonary hypertension.

Due to the complexity of the diagnosis and treatment of scleroderma lung disease strong consideration should be given to referral of patients to physicians
with expertise in scleroderma, interstitial lung disease and PAH. This requires close collaboration between you, your rheumatologist, pulmonologist and cardiologist.

Treatment of Scleroderma Lung Disease – This is very much dependent on the type of lung disease as determined by the outcome of the studies described above. For example, interstitial lung disease might be treated with immunosuppressive drugs, e.g. cyclophosphamide, azathioprine, or mycophenolate mofetil, depending upon the stage and degree of activity of the pulmonary fibrosis. Pulmonary hypertension requires different treatments, and there are a number of oral, inhaled and intravenous medications now available to treat this complication of scleroderma. Other pulmonary complications might require different treatment approaches. Adjunctive therapy is appropriate for scleroderma patients regardless of the particular type of lung disease. Adjunctive treatment might include one or more of the following: (1) avoid tobacco exposure; (2) take steps to minimize acid reflux; (3) exercise appropriately; and (4) use supplemental oxygen when prescribed by your physician.

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DANSK oversættelse

Lungesygdom ved sklerodermi : Hvad patienten bør vide

Lungesygdom kan være kompliceret i sklerodermi. De 2 mest almindelige typer ved sklerodermi patienter er interstitiel lungesygdom, også kaldet pulmonal fibrose, og pulmonal hypertension. Disse lungeproblemer kan forekomme hos patienter med limiteret og diffus sklerodermi. Selvom disse er de mest almindelige manifestationer, findes der andre mere sjældne forekomster af lungesygdomme ved sklerodermi. Se tavlen nedenfor

Typer af sklerodermi lunge sygdomme

· Interstitiel lungesygdom
· Pulmonal hypertension
· Luftvejsobstruktion
· Aspiration
· Extrinsic begrænsning
· Lungekræft
· Muskelsvaghed
· Pleural effusion
· Lungebetændelse
· Pneumothorax
· Pulmonal Blødning
· Sarcoidosis

Vurdering af sklerodermi lungesygdomme

Det er vigtigt at nævne at patienter kan have signifikante lunge involvering fra deres sklerodermi før symptomer opstår. DERFOR ER DET VIGTIGT AT FORETAGE REGELMÆSSIGE UNDERSØGELSER FOR MULIG LUNGEINVOLVERING, ISÆR INTERSTIEL LUNGESYGDOM OG PULMONAL HYPERTENSION.

Symptomer:

Brystsmerter, især ved anstrengelse, er et potentielt alvorligt symptom og bør vurderes hurtigt af din læge. Brystsmerter kan være et advarselssignal om, at du er i risiko for at få et hjerteanfald. Brystsmerter kan også forekomme hos patienter, der har pulmonal hypertension, eller det kan være et symptom på gastroøsofageal refluks (GERD).

Hoste kan ses hos patienter med interstitiel lungesygdom, pulmonal hypertension eller med luftveje sygdom. Men den mest almindelige årsag til kronisk hoste er gastroøsofageal refluks, hvilket er meget almindeligt hos patienter med sklerodermi. Det er vigtigt at bemærke, at mange patienter med GERD-relaterede hoste ikke har typiske symptomer på GERD såsom fordøjelsesbesvær eller halsbrand. Typisk i sklerodermi patienter hoste er ikke-produktiv.

Uklarhed eller besvimelse (synkope) ved fysisk anstrengelse er også et meget alvorligt symptom, der bør få umiddelbar vurdering af Deres læge. Det kan være et resultat af pulmonal hypertension eller en konsekvens af for lidt ilt sekundære til svær interstitiel lungesygdom.

Muskelsvaghed kan ses hos patienter med sklerodermi, især i de patienter, som har en overlapning med polymyositis og dermatomyositis. Patienter med muskelsvaghed i arme og ben kan også have muskelsvaghed af respirationsmusklerne.

Palpitationer (hjerte væddeløb eller flagrende) kan være et symptom på pulmonal hypertension eller hjertesygdom, og bør vurderes hurtigt af Deres læge

Åndenød, også kaldet dyspnø, er det mest almindelige symptom hos patienter med sklerodermi lungesygdom og bør straks vurderes af Deres læge. Dog kan patienter har signifikant pulmonal påvirkning uden at dette symptom. Ofte patienterne gradvist aftage deres aktivitet for at undgå den ubehagelige følelse af åndenød, uden selv at vide det. Det er vigtigt at screene for sklerodermi lungesygdom, selvom du ikke har åndenød.

Hævelse er også kaldet ødem. Nedre ende hævelse er en potentielt alvorlig symptom og bør vurderes hurtigt af Deres læge. Hævelse kan forekomme hos patienter med pulmonal hypertension, meget avanceret interstitiel lungesygdom og hos patienter med venstre-sidet hjertesygdom (hjerteinsufficiens). Visse medikamenter kan også forårsage ben ødem.

Hvæsende vejrtrækning er ikke almindeligt i sklerodermi, men kan ses hos patienter med sklerodermi-relaterede luftveje sygdom, eller hos patienter med en anamnese med astma eller rygning. Airways sygdom er mere almindelig i sklerodermi patienter, der også har træk af leddegigt eller Sjögrens syndrom. Hvæsen kan også være et symptom på gastroøsofageal refluks.

Tests din læge bør bestille

Test af lungefunktionen
Test af lungefunktionen (PFTs) udføres ved at blæse i en slange, der er forbundet til et edb-maskine.
Det er vigtigt, at du danner en forsegling med mundstykket. Ofte patienter med sklerodermi nødvendigt at bruge et talerør lavet for børn, fordi de ikke kan åbne deres mund meget bredt.

Talrige målinger kan foretages i løbet af lungefunktionen test og kan tyde på en specifik diagnose.

6 minutter walk test (6MWT) er enkel og reproducerbar og korrelerer med resultaterne på mere formel øvelse test. Det har også vist sig at korrelere med hemodynamics målt ved hjertet kateterisation, funktionelle klassificering, prognose og endda overlevelse. Den 6MWT rutinemæssigt anvendes i studier af pulmonal hypertension. I løbet af denne test, bliver du bedt om at gå så langt som du kan i løbet af seks minutter. Du har lov til at hvile, hvis du har brug for.Mens gør denne test, din puls, blodtryk, ilt niveau, graden af åndenød og andre symptomer er overvåget. Spørg din læge til at bruge en pande sonde til at overvåge din ilt niveau, som det ofte er vanskeligt at påvise ilt niveauer med en finger sonde hos patienter med Raynaud fænomen.

Tabel: Anbefalinger for lungefunktionen og Exercise Testing
* Der er ingen officielle anbefalinger for lungefunktionen og motion forsøg hos patienter med sklerodermi. Men mange eksperter anbefaler følgende:
• Baseline komplet sæt af test af lungefunktionen (spirometri, lunge mængder, diffusion)
• £ 5 års Scleroderma
- PFTs og 6MWT hver 3-6 måneder

•> 5 år Scleroderma
- PFTs og 6MWT hver 6-12 måneder

Bryst radiografi (røntgen af thorax) er ikke en følsom test for sklerodermi lungesygdom. Det betyder, at du kan have betydelige lungesygdom og stadig have en normal røntgen af thorax.Men der er træk på røntgen af thorax, der bør få din læge til at gøre yderligere forsøg.

Brystet CT-scanning (CT) er en særlig type af x-ray, der tillader en at se flere detaljer i lungerne.Under en CT, ligger du på ryggen og glide gennem en maskine, der ligner en kæmpe doughnut.Du kan også blive bedt om også at ligge på maven (udsat). Billederne er typisk taget mens du holder vejret i inspiration. Dog kan billederne også blive truffet i løbet udløb for at se, om luften er fanget i lungerne. Endelig kan du få kontrast farve for at få et bedre kig på blodkarrene i lungerne og til at lede efter tegn på blodpropper. En høj opløsning brystet CT (HRCT) (eller tynd-slice CT) betragtes som den gyldne standard for evaluering af interstitiel lungesygdom. Fund, der tyder på interstitiel lungesygdom omfatte slib opaciteter (også kaldet "alveolitis"), som ligner et haziness over lungerne, septumdefekter fortykkelse, som er ekstra linjer i lungerne på grund af fortykkelse af interstitium og honeycombing, som repræsenterer huller i lungerne. Slib opaciteter der tyder på inflammation i lungerne, som kan være reversibel, mens honeycombing udgør sædvanligvis irreversibel fibrose.

Bronchoalveolar lavage (BAL) ved bronkoskopi er undertiden gjort for at lede efter betændelse eller tegn på infektion i lungerne. En fiberoptiske omfang føres gennem næsen eller munden ned i dine luftveje. Små mængder af sterilt saltvand derefter injiceres i et område af din lunge-og suctioned tilbage ud og sendes til laboratorieanalyse. Selv Bronkoskopi og BAL til at kigge efter infektion er en rutinemæssig procedure, analysen af den væske, for forekomst af inflammatoriske celler (alveolitis), der skal gøres af erfarent personale, som regel på et sklerodermi eller pulmonal henvisning center.

*** Der er ingen officielle anbefalinger for ekkokardiogram forsøg hos patienter med sklerodermi.Men mange eksperter anbefaler følgende:**

• Baseline og årlig screening ekkokardiogram
• Patienter med pulmonal hypertension
- Ekkokardiogram hver 3-6 måneder
- Ekkokardiogram hvis forringelse i klinisk status

Ekkokardiogram er faktisk en ultralydsundersøgelse af hjertet. Ved at gnide en sonde på tværs af brystet, den kardiolog og kan se på, hvor godt dit hjerte at pumpe, se om din hjerteklapper er i arbejde, vurdere størrelsen af de forskellige kamre i hjertet, se, om der er væske omkring hjertet (perikardieeffusion), og vurdere din lungepulsåren pres.

Ekkokardiogram

Heart kateterisation

· **Højre hjerte** kateterisation er afgørende for vurderingen af pulmonal hypertension. Den hjertespecialist vil tråd en tråd gennem enten din collum vene i din lyske eller interne halsfedt i halsen til højre side af dit hjerte og i din lungepulsåren for at måle trykket i højre side af hjertet og i lungerne . Den rigtige hjerte kateteret kan også bruges til at måle din minutvolumen (hvor godt dit hjerte er pumpning), og kan vurdere pres på den venstre side af hjertet. * En ret hjerte kateterisation skal gøres af en erfaren kardiolog eller pulmonologist før du starter medicin for pulmonal hypertension.

· **Venstre hjerte** kateterisation er normalt udføres af gevindskæring en ledning gennem din femoral arterie i din lyske. Den hjertespecialist vil gøre en venstre hjerte kateterisation, hvis han / hun er på udkig efter blokeringer i din koronararterierne, eller behov for at måle tryk på venstre side af hjertet i forbindelse med pres på højre side af hjertet. En venstre sidet hjerte kateterisation er også undertiden nødvendig hos patienter med hjerteklap problemer.

Ventilation / Perfusion Scan (V / Q-scanning) er en speciel nukleare x-ray at se, om områder af din lunge at få luft også får blod. En manglende eller defekt foreslår en blodprop. Denne test bør overvejes hos patienter med pulmonal hypertension, fordi hvis blodpropper i lungerne er identificeret, kan de undertiden fjernes operativt, og pulmonal hypertension helbredt.

Lunge Biopsi er sjældent indiceret hos patienter med sklerodermi og er især risikabelt, hvis du har pulmonal hypertension.

**** På grund af kompleksiteten af diagnosticering og behandling af sklerodermi lungesygdom stærke bør overvejes at henvisning af patienter til læger med ekspertise i sklerodermi, interstitiel lungesygdom og PAH. Dette kræver et tæt samarbejde mellem dig, din reumatolog, pulmonologist og hjertespecialist.***

Behandling af Scleroderma lungesygdom - Dette er meget afhængig af hvilken type lungesygdom, som bestemmes af resultatet af de undersøgelser, der er beskrevet ovenfor. For eksempel kan interstitiel lungesygdom behandles med immunsuppressive lægemidler, fxcyclophosphamid, azathioprin, eller mycophenolatmofetil, afhængigt af tidspunkt og graden af aktivitet i pulmonal fibrose. Pulmonal hypertension kræver forskellige behandlinger, og der er en række mundtlige, inhalerede og intravenøs medicin nu tilgængelig til behandling af denne komplikation af sklerodermi. Andre pulmonale komplikationer kan kræve forskellige behandlingsmetoder. Adjuverende behandling er egnet til sklerodermi patienter uanset den særlige type lungesygdom. Adjuverende behandling kan omfatte en eller flere af følgende: (1) undgå tobak eksponering (2) tage skridt til at minimere sure opstød (3) udøve passende, og (4) anvender supplerende ilt, hvis de er ordineret af Deres læge.

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