June 16, 2017

Part B Lung Diseases Subcommittee

Advisory Board on Toxic Substances and Worker Health

DRAFT Responses to

Comments and Questions from the DOL April, 2016 Meeting

and Recommendations related to Part B Lung Disease

This document contains the responses of The Advisory Board (Part B Lung Diseases Subcommittee) to the DOL's Comments and Questions, and 3 specific recommendations and rationale. The comments also address many of the concerns regarding Part B claims that workers and others have expressed to the Advisory Board. The comments, recommendations and rationale are based on the following: The Subcommittee's analysis of data on Part B cases; a review of approximately 80 Part B case adjudications (selected records on); a review of relevant sections of EEOICPA and the 2004 Part E Addendum; relevant DOL Procedure Manuals, Circulars, and training materials; Advisory Board meetings to date and comments submitted to the Advisory Board; visits to Oak Ridge, TN and Hanford, WA sites; the relevant published medical literature, and our collective expertise.

SPECIFIC RECOMMENDATIONS RELATED TO PART B

Recommendation 1) The Advisory Board recommends that the finding of two borderline BeLPT tests shall be considered the equivalent of one positive BeLPT for the purposes of claims adjudication under subpart B and subpart E of EEOICPA. *(Voted on April 2017).*

Rationale: Beryllium sensitization is defined by EEOICPA as a single abnormal BeLPT, most commonly performed on fresh peripheral blood cells. The BeLPT result is occasionally reported as "borderline." Two "borderline" BeLPT tests have been shown to give about the same predictive value for beryllium sensitization as one abnormal (positive) BeLPT test. Thus a person with two 'borderline" BeLPT tests should be considered sensitized to beryllium (BeS), and further BeLPT testing is not indicated. The BeLPT is not a perfect test for BeS; false negative and positive BeLPT results can occur.

DRAFT Recommendation 2) The following criteria are proposed to define a clinical course consistent with a *"chronic respiratory disorder"* for use in evaluating pre-1993 CBD claims:

i) Respiratory symptoms (e.g. shortness of breath, cough) that are chronic* PLUS
ii) ONE of the following**:
- a) Abnormal pulmonary function tests (PFTs) OR
- b) Abnormal chest imaging (chest x-ray or CT scan) OR
- c) Hypoxemia, OR
- d) Chronic* use of respiratory medications such as asthma or COPD inhalers

*"Chronic" indicates symptoms (or medication usage) that are present for more than several months, to differentiate from symptoms (or medication usage) related to an acute infection or other problem that resolves.
**Not obtained during an acute illness (such as pneumonia or upper respiratory tract infection) that subsequently resolves.

Rationale: The current Procedure Manual and training materials contain several different and at times inconsistent criteria for a "Chronic respiratory disorder" for use in evaluating pre-1993 CBD claims. It is recognized that there is no one perfect criteria given the variability in clinical presentations, documentation by clinicians, access to medical care, medical record availability, and other factors. The recommended criteria are intended to provide greater consistency and ease of use.

DRAFT Recommendation 3) The Advisory Board recommends a substantial revision of sections of the Procedure Manual and related materials relevant to Part B conditions, taking into consideration the comments in this document and other feedback from the Advisory Board.

Rationale: Sections of the current Procedure Manual and related materials are inconsistent, confusing and at times medically inaccurate, which can hinder proper adjudication of Part B claims.

Endorsement: The Advisory Board endorses a presumption of chronic beryllium disease (CBD) in situations with a diagnosis of sarcoidosis in an individual who meets the definition of a "covered beryllium employee" under Part E or Part B.

Rationale: This presumption already exists, as stated in *EEOICPA Circular No. 08-07 and EEOICPA Procedure Manual Chapter 2-1000.* However, implementation of this presumption has been problematic. Revising the relevant Sections of the Procedure Manual and training materials, within the statuary limitations of EEOICPA, should help alleviate this problem.

RESPONSES TO DOL'S SPECIFIC COMMENTS AND QUESTIONS

1. Beryllium Sensitivity

DOL Comments / Questions:
1) Consistency of testing results among the different diagnostic facilities

Response:

National Jewish Medical Center (Denver, CO), Oak Ridge Associated Universities (ORAU), and the Cleveland Clinic (Cleveland, OH) are the only laboratories in the U.S. that we are aware of that currently perform the beryllium lymphocyte proliferation test (BeLPT) on a regular basis. These laboratories have extensive experience performing the BeLPT test. Consistency between these laboratories has improved and does not appear to be an on-going issue. Additional laboratories would likely increase problems with the accuracy and reproducibility of performing BeLPT testing.

2) Reinterpretation of "normal" test outcomes as abnormal by a consulting physician.

Response:

A patient's BeLPT report from the lab performing the test should not be reinterpreted by a consulting physician.

However, the quality and interpretation of the standard clinical tests used to evaluate patients with pulmonary disorders (chest x-ray and CT scans, pulmonary function testing, lung pathology), can be quite variable and significant inter-observer variability can occur[1]. These tests involve interpretation of multiple images, visual patterns, and/or data points, and treating or consulting physicians routinely re-review the studies themselves or with the appropriate specialist (e.g. chest radiologist, pulmonary pathologist). Proper interpretation also can require comparison to prior testing results if available.

3) New and more relevant science on diagnostic tools for evaluating beryllium sensitivity?

Response:

There are no additional or better diagnostic tests for BeS or CBD.

Of note, patch testing to beryllium, which involves putting a test amount of beryllium on the patient's skin, is no longer recommended to assess sensitization to beryllium, as performing the test can induce beryllium sensitization in someone not previously sensitized.[2] Skin patch testing is referred to as a possible diagnostic test for BeS throughout the EEOICPA Procedure Manual (Chpt 2-1000) and related training materials, which creates confusion and should be removed. Also tissue beryllium levels should not be used to diagnose BeS or CBD.

4) Definition of beryllium medical monitoring i.e. expected medical regimen for monitoring sensitivity to determine if it has progressed to CBD.

Response:

For those who are sensitized to beryllium the American Thoracic Society recommends periodic medical monitoring every 2 to 3 years, or sooner if there is concern about progression[2]. This evaluation should include a review of symptoms, physical examination and pulmonary function testing. If deterioration is noted, (or prior chest imaging warrants follow-up evaluation), a chest CT scan is recommended. Further evaluation, such as bronchoscopy or lung biopsy or cardiopulmonary exercise testing, is considered on a case-by-case basis.[2,3]

2. Chronic Beryllium Disease

DOL Comments / Questions:

A) Pre-1993 CBD

1) Characteristic chest radiographic or computed tomography (CT) abnormalities. More clear guidance on chest radiographic abnormalities consistent with CBD would be useful.

Response:

The radiographic abnormalities (chest x-ray and CT scan) seen with CBD are variable and non-specific. In addition to small nodules in the lung parenchyma (considered the most common finding), ground glass opacities, bronchial wall thickening, thickened interlobular septa, lymphadenopathy (hilar, mediastinal), and no radiographic abnormalities are also seen. With more advanced disease, interstitial fibrosis, honeycombing, subpleural cysts, traction bronchiectasis and calcifications can be seen.[2,4]

The chest xray and chest CT scan imaging findings that can be seen with CBD are generally described appropriately in Chapter 2-1000 (Section 6, pgs 5-6). Parts of the text are confusing and would benefit from editing. Specific suggested edits include:

Discussion regarding granulomas (Section 6 pg 5): Whether a granuloma is caseating or not is determined by pathologic examination of lung tissue and not chest imaging. This text should be removed from the chest x-ray section.

Similar to sarcoid granulomas, CBD granulomas can become calcified. Thus the sentence *"A calcified granuloma is not characteristic of CBD"* is incorrect and should be removed.

The EEOICPA statutory criteria refer to "characteristic chest x-ray or CT abnormalities". Any of the imaging findings noted in the Training manual or above that are *"consistent with"* or *"seen with"* CBD" are also *"characteristic abnormalities".*

2) Restrictive or obstructive lung physiology testing or diffusing lung capacity defect. Pulmonary function test (PFT) is used as diagnostic tool for specific illnesses (i.e. asthma, COPD). Are PFT results within certain ranges consistent with CBD?

Response:

Pulmonary function tests (spirometry, lung volumes, diffusing capacity) evaluate how well lungs work, such as airflow, lung size, ability to provide oxygen, and can help diagnose lung diseases, including CBD and COPD. Thus PFTs can identify abnormalities in how a person's lung function, but do not diagnose specific illnesses such as CBD, silicosis or COPD.

The PFT findings in CBD, similar to the findings in sarcoidosis, are variable and non-specific, and can also fall within normal limits. Thus normal lung function, airflow obstruction, restriction (reduced lung volumes), low diffusing capacity, and mixed air flow obstruction / restriction can be seen in CBD. With more advanced CBD, a reduced diffusing capacity (DLCO) and restriction are more common, but other PFT findings can be present.

Of note, it is important to compare lung function results with prior testing when available, as there can be significant declines in lung function, but values can still fall within normal ranges.[5] A recent American Thoracic Society (ATS) document provides guidance to performing and interpreting spirometry related to work exposures.[5]

3) Lung pathology consistent with CBD.

In most instances, a physician's statement with medical rationale confirming that pathologic test results are consistent with CBD is sufficient to support claim. Additional guidance on lung pathology findings consistent with CBD would be useful.

Response:

The typical lung pathology in CBD is a non-caseating granuloma that is indistinguishable from sarcoidosis. The granulomas can be well formed or a loose collection of epitheliod cells. The granulomas can be in the lung itself, in chest lymph nodes or in other organs, such as the skin, liver or nose. However, granulomas are not always seen on lung pathology.[2,6]

Other findings that are consistent with CBD include: interstitial infiltrates (lymphocytes, monocytes, plasma cells), and multinucleated giant cells. In more advanced disease, progressive

fibrotic changes can be seen, including diffuse fibrosis, fibrotic nodules, foreign body granulomas, calcifications, cystic-honeycomb changes, and bronchiectasis.[2,6] These findings are also seen in other chronic interstitial lung diseases, such as sarcoidosis, chronic hypersensitivity pneumonitis, and idiopathic pulmonary fibrosis.

The text in the Procedure Manual Chpt 2-1000 regarding pathology contains some inaccuracies and requires revision. For example:

"a mediastinal lymph node biopsy interpreted by a physician as evidence of "lung pathology consistent with CBD" may be used to establish CBD. A mediastinal lymph node biopsy is not the equivalent of a "lung biopsy" and, as such, does not substitute for such in the assessment of a post-1993 CBD claim. The evidence has to be interpreted as "lung pathology." A mediastinal lymph node is not dispositive proof of CBD in the same way as a lung biopsy." (Section 7 Post-1993 Criteria)

Comment: A mediastinal (or any chest) lymph note biopsy showing findings consistent with CBD IS the equivalent of a lung biopsy and CAN be substituted for a lung biopsy. Lymph nodes (or extrapulmonary sites) are sometimes biopsied instead of lung tissue to reduce the risk of complications such as pneumothorax. The wording above should be deleted.

"If a pathology report does not include a physician's interpretation... the CE obtains clarification from a treating physician or CMC." (Section 6f).

Comment: Pathology reports ARE a physician's description and interpretation of the findings. With non-malignant lung diseases such as CBD, sarcoidosis, or asbestosis, the pathologic findings commonly are non-specific, so reports frequently describe the pathologic findings, such as granulomatous lung inflammation, inflammatory or fibrotic changes, but do not provide a specific diagnosis such as CBD. Thus any of the pathologic findings noted above in a worker with BeS should be considered diagnostic for CBD. The pathologist's report does NOT need to state that the findings are consistent with CBD.

B. Post-1993 CBD Criteria

DOL Comments / Questions: CBD criteria are: Beryllium sensitivity AND lung pathology consistent with CBD, including lung pathology, CT scan, and PFT consistent with CBD.

Issue *1. Clarification of the diagnostic and interpretive meaning of "characteristic of CBD" to differentiate between CBD and other lung disease.*

Response:
Lung pathology, chest imaging, and PFT findings considered "consistent with CBD" are described above and should be the same for Pre-CBD and Post-CBD diagnostic criteria. As noted, these test results are not unique to CBD, and it is not possible to differentiate CBD from sarcoidosis, discussed further below.

Issue 2. Consistent and uniform standard for judging medical evidence for the pre or post 1993 as evidence of a "chronic respiratory disorder".

Response:

A consistent standard should be used to define a "*chronic respiratory disorder"* for Part B lung conditions. A "chronic respiratory disorder" is one of the diagnostic criteria for pre-1993 CBD, not post 1993 CBD. Thus the proposed standard below is relevant only for pre-1993 CBD diagnoses.

Chronic respiratory disorders such as CBD or COPD commonly are present and progress slowly for years before they come to the medical attention especially when present in those who are not under regular respiratory medical surveillance. Chronic respiratory conditions also frequently are initially recognized after an acute infection or event and may initially be misdiagnosed as an acute illness such as bronchitis or pneumonia.

A consistent standard for judging whether medical evidence meets the pre 1993 conditions for *"chronic respiratory disorder"* is the following:

i) Respiratory symptoms (e.g. shortness of breath, cough) that are chronic* PLUS
ii) ONE of the following**:
- a) Abnormal pulmonary function tests (PFTs) OR
- b) Abnormal chest imaging (chest x-ray or CT scan) OR
- c) Hypoxemia, OR
- d) Chronic use of respiratory medications such as asthma or COPD inhalers

The criteria for abnormal PFT or chest imaging are as described above.

*Chronic typically means persistent, present for several months duration, to differentiate from symptoms (or medication usage) related to an acute infection or other acute problem that resolves.

**Not obtained during an acute respiratory illness such as pneumonia or upper respiratory tract infection that subsequently resolves.

The Procedure manual Chpt 2-1000 and training materials at times provide inconsistent and/ or inaccurate descriptions of a "*chronic respiratory disorder"* and require revision. For example:

1) Whether to use the pre- or post- 1993 CBD criteria depends upon the totality of the medical evidence, including when the employee was tested for, diagnosed with, and/or treated for a chronic respiratory disorder. (Section 6)

2) If the earliest dated document showing that the employee was either treated for or diagnosed with a chronic respiratory disorder is dated prior to January 1, 1993, the pre-1993 CBD criteria should be used. Evidence of a chronic respiratory disorder includes records communicating existence of a long-term, prolonged pulmonary disease process. References to acute pulmonary conditions, such as short-term pulmonary distress associated with temporary viral or bacterial infection do not qualify as a chronic respiratory disorder. Pulmonary testing performed in occupational or medical settings, which identify abnormalities, are not appropriate to document a chronic respiratory disorder, unless interpreted as such .... (Section 6; CBD Pre 1993)

Comment: When a patient is "*tested, treated for or diagnosed"* can be very different time periods; the terms should be used consistently. As noted, chronic respiratory conditions may be present for years before they come to medical attention and are diagnosed as a chronic respiratory condition. They also may initially be diagnosed as an infection or acute bronchitis. Pulmonary function tests are generally not performed when patients are acutely ill (per ATS guidelines).[5] Thus abnormalities noted on PFTs (obstructive, restrictive, reduced DLCO) that are performed properly in either an occupational or medical setting generally DO indicate a chronic respiratory disorder. Of note, per ATS guidelines, interpretations on PFTs reports describe the physiologic abnormalities present but do not state whether such findings document a chronic respiratory disorder such as CBD or COPD.[7]

3) Clinical course consistent with chronic respiratory disorder may include the following .....:
(1) Hypoxemia requires supplemental oxygen...
(2) Air flow obstruction (e.g., COPD ...) and asthma/wheezing--like symptoms require inhalers ....
(3) Right heart failure, Cor pulmonale....
(4) Pulmonary Hypertension ...
(5) Respiratory infections (pneumonia, acute bronchitis) ...
(6) Sarcoidosis: corticosteroid drugs ...
(Section 6g CBD Pre-1993 Criteria)

Comment: The text above is not consistent with other parts of Chpt 2-1000 and / or medical guidelines. For example, patients can have sarcoidosis but not be on corticosteroids.

Replacing the current multiple criteria for a “chronic respiratory condition” in the Procedure manual Chpt 2-1000 and training materials with a consistent standard for pre 1993 CBD such as the one noted above should facilitate implementation of EEOICPA.

Issue 3. Necessitating lung lavages or lung biopsy on critically ill or elderly patients

Response:

Bronchoscopy with lung lavage and/or lung biopsy is generally contraindicated in critically ill or elderly patients due to the risks of the procedures, which include bleeding, intubation and ventilation, pneumothorax and infection.[8,9]

Issue 4. Obtaining clarity on the specific diagnostic markers required for CBD in the pre or post 1993 diagnostic requirements.

Response:

As noted above, there is no one single diagnostic test for CBD and no specific diagnostic markers “required for CBD”. There are also no new or additional diagnostic tests available to diagnose CBD. The text of Chapter 2-1000 regarding pre and post 1993 CBD diagnostic criteria would benefit from revision and greater consistency as noted above and below.

Issue 5. Clearer guidance on the relationship between sarcoidosis and CBD

Response:

The Advisory Board endorses a presumption of chronic beryllium disease (CBD) in situations with a diagnosis of sarcoidosis in an individual who meets the definition of a “covered beryllium employee” under Part E or Part B. This presumption already exists, as stated in *EEOICPA Circular No. 08-07 and EEOICPA Procedure Manual Chapter 2-1000.* However, implementation of this presumption has been problematic. Revising the current text of the relevant Sections of the Procedure Manual (Chpt 2-1000) and training materials, while remaining within the statuary limitations of EEOICPA, should alleviate this problem.

Several key features regarding sarcoidosis and CBD should be clarified:

Sarcoidosis is as a multisystem granulomatous disorder that clinically and pathologically is difficult to differentiate from CBD.[10-12] The granulomas seen histologically in both conditions are indistinguishable and both can have extrapulmonary involvement. Sarcoidosis predominantly affects the lungs and thoracic (hilar and mediastinal) lymph nodes, but can also involve other organs including the skin, nose, liver, heart, other lymph nodes and the musculoskeletal system. About 90% of sarcoidosis patients have pulmonary involvement.[13,14]

The diagnosis of sarcoidosis is typically based on a tissue biopsy that shows non-caseating granulomas in one of the affected organs (such as skin, nose, liver, lymph nodes, lung) along with the patient’s clinical presentation and other diagnostic tests.[13,15,16] The organ biopsied (e.g. skin, nose) usually indicates the safest way to obtain diagnostic tissue, not what organs are involved. Thus pulmonary sarcoidosis not uncommonly is diagnosed by biopsy of an organ such as skin or lymph nodes, rather than lung tissue. In such cases lung involvement is usually confirmed by chest imaging and/or pulmonary function testing.

Published articles may highlight certain features that are more common in sarcoidosis vs. CBD.[10,11] For example, extra pulmonary involvement is more common in sarcoidosis than CBD, and the prevalence of sarcoidosis is higher in blacks than Caucasians. However, extrapulmonary involvement occurs in both sarcoidosis and CBD, and both diseases occur in all racial groups. Thus such features should not be used to differentiate CBD and sarcoidosis in an individual.

Chronic sarcoidosis is an uncommon condition, with a prevalence in the USA estimated at about 50 -150 per 100,000[17]. Sarcoidosis is much less common than CBD in Be-exposed workers, which has a prevalence estimated to range from about 0.1%-7% (0.1 - 7 per 100).[2,18]

An abnormal BeLPT test, documentation of BeS, is the most definitive way to confirm CBD in a patient that has been diagnosed with sarcoidosis. However, there are a number of reasons why a worker with CBD may not have a documented abnormal BeLPT test and be misdiagnosed as having sarcoidosis.

Steroids and other immunosuppressive medications used to treat CBD, sarcoidosis and other chronic lung conditions can suppress the proliferation response of blood and /or lung lymphocytes to beryllium, resulting in a false negative BeLPT. This is currently noted in the Procedure Manual Chpt 2-1000: *"If the claimant has a history of steroid use, a false negative BeLPT .... can occur".* Section 5c (Beryllium Sensitization; False negative results).

False negative BeLPT tests have also been documented in patients even if not on immune-suppressive medications.[19-23,24] The BeLPT test performed on lung lavage cells, while more sensitive than the peripheral blood BeLPT can also have false negatives due to smoking, immunosuppressive treatment and other factors.[25]

The BeLPT is not a routine clinical test and is rarely performed, except in the fraction of beryllium-exposed workers enrolled in a beryllium medical surveillance program. Thus it is common for covered beryllium employees, despite outreach efforts by DOE, DOL and others, not to have had a BeLPT test performed. This is currently noted in the Procedure Manual Chpt 2-1000:
"If exhaustive efforts produce little or no results (BeLPT testing) ..... the CE can accept the claim.
(Section 5c).

Our review of Part B lung CBD and sarcoidosis decisions found cases of CBD hat had been incorrectly adjudicated and denied. Some cases had BeLPT tests that were likely false negatives. Other employees had never had a BeLPT test performed, either because the employee had not been in a beryllium surveillance program and/or the provider (including at DOE sites) did not consider CBD and/or never obtained the BeLPT test.

Our review of cases also demonstrated that most clinicians, even board-certified pulmonary and occupational medicine physicians, know little about CBD, BeS, BeLPT testing, or their patient's work history. This unfortunately was true for some current CMCs, despite the current Procedure Manual and training materials.

In summary, in situations where a "covered beryllium employee" under Part B or E has been diagnosed with pulmonary sarcoidosis, that employee is much more likely to have CBD than sarcoidosis and should be diagnosed with CBD, even if the BeLPT is normal (may be false negative test) or was never performed.

The current Procedure Manual and EEOICPA Circular 08-07 similarly state that sarcoidosis is not an appropriate diagnosis in a covered beryllium employee and explain how to accept a claim for CBD in this setting, including when the BeLPT is normal or was never performed.

However, the current wording in the Procedure Manual Chpt 2-1000 (Section 10) and related materials is confusing, inconsistent and requires substantial revision. For example:

"Presumption of CBD, Diagnosis of Sarcoidosis, and History of Beryllium Exposure. ....."Under Part B, the DEEOIC recognizes that a diagnosis of pulmonary sarcoidosis, especially in cases with pre--1993 diagnosis dates, could represent a misdiagnosis for CBD. As such, a diagnosis of pulmonary sarcoidosis is not medically appropriate under Part B if there is a documented history of beryllium exposure..... Under Part E, if there is a diagnosis of pulmonary sarcoidosis, but no affirmative evidence in the form of a positive BeLPT or BeLTT exists, the CE adjudicates the condition as sarcoidosis, not CBD."

Comment: A diagnosis of pulmonary sarcoidosis most likely (not *"could")* represents a misdiagnosis of CBD. This is the case whether or not the CBD claim is made pre or post 1993, the BeLPT is positive or negative, or the employee is applying under Part B or Part E.

Alternate text for Chapter 2-1000 Section 10 that meets the statutory criteria of EEOICPA Parts B and E and can be used to describe how to diagnose CBD in a covered beryllium employee with sarcoidosis is as follows:

"*A diagnosis of pulmonary sarcoidosis is not medically appropriate in a covered beryllium employee under Part B or Part E as the employee most likely has CBD that has been misdiagnosed as sarcoidosis. In these situations, the CE is to consider the diagnosis of sarcoidosis to be a diagnosis of CBD. The following should be documented to determine this diagnosis:*

a) When sarcoidosis is diagnosed in a "covered beryllium employee" under Part B or Part E, pulmonary sarcoidosis should be documented as noted above. The presence of granulomas (=granulomatous inflammation) on tissue biopsy should be documented. The tissue where granulomas are identified can be extrapulmonary, such as skin, nose, or lymph nodes. In such cases pulmonary involvement, which is present in over 90-95% of those with sarcoidosis, can be documented based on PFT or chest imaging findings, as noted above.[15,26]

b) The results of any available BeLPT testing should be documented. If the BeLPT test is normal (negative for BeS), reasons for a false negative BeLPT (such as immunosuppressive treatment or recruitment of sensitized lymphocytes to the lung) should be documented, as noted above. Repeat BeLPT testing is not indicated when there is a diagnosis of pulmonary sarcoidosis in a covered beryllium worker, as the appropriate diagnosis is CBD whether or not the repeat BeLPT testing is abnormal .

If BeLPT testing was not performed, the reason why should be noted, such as the worker is deceased or was not in a beryllium medical surveillance program."

<u>Rationale for using the same CBD / sarcoidosis presumption criteria under Part B and E:</u>

The current wording regarding adjudicating CBD / sarcoidosis claims under Part B and E is confusing. (Procedure Manual: Chpt 2-1000 Section 10).

The statutory requirements of EEOICPA Part E require that the Secretary of Labor of determine that *'it is at least as likely as not that exposure was a significant factor in aggravating, contributing to, or causing the illness".* The Procedure Manual states for Part E claims: *"In cases where there is .... a diagnosis of pulmonary sarcoidosis and a positive BeLPT, the CE is to obtain a physician's opinion regarding whether it is 'at least as likely as not' that exposure was a significant factor in aggravating, contributing to, or causing CBD".* In CBD / sarcoidosis cases this Part E statutory requirement has already been met and exceeded. A physician's opinion is not needed
This requirement can be eliminated.

Our understanding is that a claim accepted under Part B as CBD will also be accepted under Part E, and if a claim for CBD is denied under Part B, it will also be denied under Part E. Thus, although the wording of EEOICPA Part B and E differ, different criteria for CBD / sarcoidosis under Part B and Part E appears to be confusing.

A claim for CBD that is denied under Part B can be accepted under Part E as another chronic lung condition, such as pneumoconiosis, according to the statutory requirements of Part E. Relevant to Part E claims more broadly, *"At least as likely or not"* and *"significant factor in aggravating or contributing to an illness"* should be defined, as many clinicians, including CMCs based on our review of cases, are not familiar with the terminology.

For clarification, the "presumption of CBD when sarcoidosis" discussion above relates to beryllium employees diagnosed with sarcoidosis. If a worker has evidence of lung disease and/or BeS but has NOT been diagnosed with sarcoidosis, then the Part B CBD criteria should be used to document a diagnosis of CBD and the Part E criteria used for other chronic respiratory conditions.

6. Recommendations or advice relating to conditions that are normal and usual consequential illnesses to CBD

Response:

The March 2016 update of Chapter 2-1500 Consequential Conditions, Exhibit 1 contains a list of disorders secondary to CBD and to CBD treatment (steroids). The list includes pulmonary hypertension, right heart failure, respiratory infections, GERD, decreased bone density, diabetes and osteoporosis. The Advisory Board agrees with the use of this list of conditions.

7. Input or suggestion regarding assessment of negative BeLPT as either false negative or borderline due to drug interference or other treatment modalities

Response:

Beryllium sensitization is defined by EEOICPA as a single abnormal BeLPT, which most commonly is performed on fresh peripheral blood cells.

The BeLPT result is occasionally reported as "borderline". Two "borderline" BeLPT tests have been shown to give about the same predictive value for beryllium sensitization as 1 abnormal (positive) BeLPT test.[27,28] **Thus a person with two 'borderline" BeLPT tests should be considered sensitized to beryllium (BeS), and further BeLPT testing is not indicated.**

The BeLPT is not a perfect test for BeS. False negative and positive BeLPT results can occur for several reasons, as noted above.[29]

3. Chronic silicosis

Comments / Questions:

1. Clear guidance on the certification requirements for B-readers and how that is documented on B-reader test results.

Response:

NIOSH provides a list of all certified B readers and also describes the B-reader certification process on its website. https://wwwn.cdc.gov/niosh-rhd/cwhsp/ReaderList.aspx. A claims examiner (and others) can verify that a B reader is currently certified by checking the NIOSH website.

B-reading of chest x-rays remains important for medical surveillance of exposed workers and can be used to diagnose pneumoconioses, but are not required by EEOICPA. Chest CT scans are now commonly used to evaluate individual patients with chronic lung diseases such as CBD, silicosis, and pneumoconiosis.

OTHER COMMENTS:

1) Chapter 2-1000 of the Procedure Manual likely has undergone several revisions. Its current wording is confusing, contains inaccuracies in certain sections and would benefit from a major revision and streamlining. Much of the inaccurate and/or confusing information, such as the discussion of calcified granulomas or the significance of granulomas in chest lymph nodes vs. lung tissue, is NOT part of the statutory requirements of EEOICPA (or the 2004 Part E Amendment) and should be removed. The Part B subcommittee would be willing to assist the DOL in revising the Procedure Manual.

2) As noted above, most pulmonary and occupational medicine physicians, including likely many CMCs, do not have experience diagnosing occupational lung diseases such as CBD. Our review of 80 Part B case adjudications supports these concerns. Revising and updating the Procedure manual is warranted but does not address issues related to the quality and oversight of the CMCs and claims examiners, and other components of the EEOICPA claims adjudication process.

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Chronic Beryllium Disease and Chronic Silicosis under the EEOICPA statute:

§ 7384l. Definitions for program administration

(13) The term "established chronic beryllium disease" means chronic beryllium disease as established by the following:

(A) For diagnoses on or after January 1, 1993, beryllium sensitivity (as established in accordance with paragraph (8)(A)), together with lung pathology consistent with chronic beryllium disease, including—

(i) a lung biopsy showing granulomas or a lymphocytic process consistent with chronic beryllium disease;

(ii) a computerized axial tomography scan showing changes consistent with chronic beryllium disease; or

(iii) pulmonary function or exercise testing showing pulmonary deficits consistent with chronic beryllium disease.

(B) For diagnoses before January 1, 1993, the presence of—

(i) occupational or environmental history, or epidemiologic evidence of beryllium exposure; and

(ii) any three of the following criteria:

(I) Characteristic chest radiographic (or computed tomography (CT)) abnormalities.

(II) Restrictive or obstructive lung physiology testing or diffusing lung capacity defect.

(III) Lung pathology consistent with chronic beryllium disease.

(IV) Clinical course consistent with a chronic respiratory disorder.

(V) Immunologic tests showing beryllium sensitivity (skin patch test or beryllium blood test preferred).

EEPICPA Part E Addendum 2004

Under Part E of the EEOICPA, benefits may be extended to DOE contractor or subcontractor employees (or their eligible survivors) whose exposure to a toxic substance at a covered DOE facility was ***"at least as likely as not"*** **a significant factor in** ***aggravating, contributing to or causing the illness.***