CENTERS FOR MEDICARE & MEDICAID SERVICES

Moderator: Donna Pickett
September 16, 2010
8:00 a.m. CT

Operator: Good morning. My name is Lisa and I will be your conference operator today. At this time, I would like to welcome everyone to the ICD-9-CM Coordination and Maintenance Committee Conference Call.

All lines have been placed on mute to prevent any background noise. After the speaker's remarks, there will be a question-and-answer session. If you would like to ask a question during this time, simply press star and the number one on your telephone keypad. If you would like to withdraw your question, press the pound key. Thank you.

Ms. Donna Pickett, you may begin your conference.

Donna Pickett: Thank you, Lisa. Good morning, everyone. Welcome to the second day of the ICD-9-CM Coordination and Maintenance Committee Meeting. We'll be reviewing diagnosis proposals. For those of you who are interested in the procedure proposals and were not able to attend yesterday, that information is available on the NC – CMS Web site, sorry Pat.

Also, referencing yesterday's meeting, for those of you who are also interested in the information about the partial freeze of the ICD-9-CM and ICD-10 code sets and any of the materials related to the General Equivalence Mappings, that information is also available in the CMS topic package. The discussion – the information on the GEMs appears on Pages 11 through 20 roughly, and on the MS-DRGs on Pages 21 to 32.

Also, for those of you who were not here yesterday or not online, please note that some of the diagnosis proposals were presented yesterday and I will review the names of those topics for you so that you can keep track of those.

Today, we will be discussing diagnosis proposals, as many of you know. But for those who maybe participating for the first time, no decisions are made during this meeting. We encourage – excuse me – we encourage your discussions and comments and questions during the meeting, but we also encourage you to send us your comments in writing by the deadlines that are noted in the CMS and NCHS topic package.

By way of summary, for comments related to the general equivalence map, those comments, the deadline is November 12th. For comments on the diagnosis proposals and as well as for the procedure proposals, those comments are due by November 19. For those of you who may be thinking about submitting new proposals for the diagnosis or procedure topics, the deadline is January 7, 2011, obviously.

A little housekeeping thing. Let me mention that now. Some of you may have seen signs – for those of you who are here in the audience, you may have seen signs about a walk-run event today.

That also includes a healthy barbeque lunch. You are able to I understand – buy tickets for that healthy barbeque lunch in the cafeteria or, of course, you can avail yourself of the regular cafeteria services. So, we hope you enjoy either/or but not both.

We will have the call-in lines available today until 3:15 p.m., but as many of you can tell, we have a lot of topics to consider during today's meeting, even though we were able to present some of them yesterday.

Even after the phone lines are closed, we will continue to meet until we get through all of the proposals. NCHS will post a summary, as we always do, of the discussions here at the meeting should we not be able to finish everything by 3:15 this afternoon.

Keep in mind, though, I probably will be a bit of a taskmaster to make sure that we try to get through as many proposals as we possibly can.

Also, because the agenda is full and similar to yesterday, we will have no formal breaks. So, just so you know, we're going to continue to keep going until lunchtime and the lunch break will be 12:30 to 1:30 today.

For those of you who weren't here yesterday, I just want to give you the names of the topics that were presented yesterday afternoon, so that if you see that they're not being presented, there is a good reason. It's not that we overlooked them.

The things that we presented yesterday were the proposals on the complications of stem cell transplant, pseudobulbar affect, dementia with or without behavioral disturbance. There were few a orthopedic issues and we also presented the addenda and also the topic, arteriovenous malformation.

And with that, I will now turn the podium over to Beth Fisher who will introduce our first topic and our online speaker.

Beth Fisher: OK. First, I'm going to ask is Dr. Sage Claydon- are you on the telephone? If you are in, unmute your phone and you can say hello, hopefully.

Dr. Sage Claydon: Hello.

Beth Fisher: Is this Dr. Claydon?

Dr. Sage Claydon: It is. Thank you for reminding me to unmute it. I picked up the phone, I'm like no one can hear me?

Beth Fisher: OK. OK. Let me just get the topic started and I'll ask you to speak in a moment. Thank you.

Dr. Sage Claydon: OK.

Beth Fisher: For those following along in the topic packet, we're on Page 16.

We aren't necessarily going to be presenting the topics in the order they appear in the topic packet, so I'll try to remind you of the page number.

The topic is mesh erosion, mesh exposure. And if you were at our meeting a year ago, September '09, it says, we did present this topic. It's from the American College of Obstetricians and Gynecologists, ACOG, and they asked us to consider a new code or new codes for mesh erosion and mesh exposure.

Specifically, they were asking for codes related to vaginal mesh erosion and exposure. We presented it in a more general fashion trying to make the code usable for other types of surgeries that use the mesh that have erosion or exposure.

And we received – most comments we received were about the terms erosion and exposure. They seem to either be used interchangeably or it wasn't really clear if the definition meant the same thing different specialties.

So, we brought the proposal back today with having Dr. Claydon online who can help us a little bit with that and also to show you that we are presenting as we did last September as Option 1 which would be to locate these codes in the Complication section at Subcategory 998.8, new code 998.84 and 85 for mesh erosion and mesh exposure and also presented a second option which we wanted to make it specific to vaginal mesh or we could do both. I suppose we haven't considered that. But the second option is to locate the codes in a new subcategory in 629.3, mechanical complications of implanted vaginal mesh. And with the code 629.31 for the erosion of implanted vaginal mesh or exposure of the implanted vaginal mesh.

But first, maybe I could just ask Dr. Claydon. Could you – would you mind giving us just an overview of this, as it relates maybe specifically to your specialty and specifically touch on the terms themselves?

Dr. Sage Claydon: Sure. It seemed everyone has read the vignette which I don't have in front of me so – but the vignette that was presented related to abdominal sacral colpopexy which is a procedure where we surround the top of the vagina with a mesh and attach that to the sacrum, the anterior longitudinal ligament of the sacrum. For that particular procedure, two things can happen.

Beth Fisher: Yes. Could you just speak up a little bit …

Dr. Sage Claydon: Oh, sure. For that particular procedure, two things can happen. Can you hear me OK now?

Beth Fisher: Yes.

Dr. Sage Claydon: OK. One thing can happen is the mesh can become exposed in the vagina, such that the vagina is essentially coated with a skin so that if – with an abdominal sacral colpopexy, if the mesh is exposed in the vagina, it's open to – there's an open connection between the vagina and the presacral space or the sacrum. And in that case, we have to remove mesh and recover it.

The other thing that can happen in that case is the mesh can erode into bowel; it can erode into the bladder. And so we use the term exposure and erosion really slightly differently where exposure means we can see it and there's no skin covering it, and erosion means it's gone into something else.

When we place those same types of meshes – and these are polypropylene meshes, the same ones that the general surgery uses for hernia repair – when we place those meshes vaginally, two things can happen.

The vaginal skin wouldn't heal over the mesh, but the mesh will grow into the muscle of the vagina making it very difficult to get the mesh out so, we'll have to take the patient back to the operating room, cut the mesh out and try to reposition skin over. And we typically refer to that as mesh exposure because we can see it, the patient can feel it, it's causing discharge and dyspareunia.

When the meshes can also find their way into the bladder, into the urethra, into the rectum and – or into the muscle – the pelvic floor muscles in such a way that they're extremely difficult to remove and we typically call that mesh erosion.

Mesh erosions are associated with a whole host of problems related to having a foreign body where it does not belong. So a foreign body in the bladder, a foreign body in the bowel, a foreign body deep in the pelvic muscles

surrounded by nerves. It can cause pain, infection and a whole host of complications, and that's a difficult thing to treat.

Exposures typically cause dyspareunia which is painful intercourse for the man or the woman, vaginal discharge and annoyance and they can get bigger over time. So, these codes are to distinguish the difference. Exposures tend to not – not all the time because you know nothing is 100 percent amount of time but they tend to be something that we're seeing mesh where it should be covered, and erosion means the mesh has gone some place it doesn't belong. Does that help clarify it?

Beth Fisher: I hope so. It helps me a little bit.

Dr. Sage Claydon: OK.

Beth Fisher: I'd like to ask the audience if you have any questions for Dr. Claydon before we take comments on the actual proposal itself? Is there any questions anyone has for her? And I'd like to also ask the operator if any of our telephone participants have any questions for Dr. Claydon?

Operator: At this time, I would like to remind everyone, in order to ask a question, press star and the number one on your telephone keypad. We'll pause for a moment.

Your first question comes from Dr. Linzer. Your line is now open.

Dr. Jeffrey Linzer: I have a question on …

Beth Fisher: Go ahead, Dr. Linzer.

Dr. Jeffrey Linzer: Hello. Is the – the intention is this not to be specifically limited to GYN procedures, but this code would apply to any surgical procedure that uses a mesh?

Beth Fisher: OK. Could you say that again, Dr. Linzer? It was hard to hear you.

Dr. Jeffrey Linzer: I say the code is not limited just to GYN procedures, but would it be used for any surgical procedure that uses a mesh?

Beth Fisher: Well, that's where we've presented two options. Option 1 was to not have it limited, to have it just be mesh in general. And then Option 2 was specifically to the vaginal mesh. So we're presenting it two different ways.

Dr. Jeffrey Linzer: Well, I guess my question is if ACOG has discussed this with any of the other surgical specialties?

Beth Fisher: We did submit it to the – Donna, where is Dr. Fry? American College of Surgeons. And they had no objections to it. I mean they didn't, I guess, comment one way or the other.

Dr. Jeffrey Linzer: OK. Thank you.

Beth Fisher: OK. OK, then I'd like to ask the audience in the room here, if you have any comments about the proposals themselves, the options, any other questions. Yes?

Linda Holtzman: Linda Holtzman of Clarity Coding. First, I guess I should say that I do think it would be a nice idea to have a code specifically for erosion or exposure but I think that the proposals are somewhat problematic and it really has to do with the language.

Part of the problem is that we're accustomed to speaking of the term erosion in terms of eroding through the skin. I mean, that always been presented to us. There are numerous coding clinic articles that discussed erosion – let's say a pacemaker through the skin or erosion of some other device through the skin – so we tend to think of erosion in those terms and we've got you know 30 years of precedent – coding precedent for that. So I think it's going to be very difficult to change – you know, do a sea change …

Beth Fisher: As opposed to that being exposure?

Linda Holtzman: Right. Exactly.

Beth Fisher: OK.

Linda Holtzman: So if we do go with something like this and I'd have to think it through, but I think we need to have numerous inclusion terms or notes or something, instructional notes that clarify exactly what is meant by erosion and exactly what is meant by exposure.

Beth Fisher: Yes.

Linda Holtzman: We're also going to have to deal with questions then on how do you code skin erosion for other implanted material and that's going to wind up back with – in the 996.76.

I mean, I'm just concerned that if we go with the first option then people are going to say, "Should I code erosion of a pacemaker through the skin to one of these what's not really mesh, on the other hand it's erosion, you know?" So it's still I think going to be fairly confusing.

Beth Fisher: OK. So we need to look a little more at our inclusion terms, indexing. Think it through a little – OK. Thank you.

Dr. Sage Claydon: May I comment on that at all?

Beth Fisher: Sure, Dr. Claydon. Go ahead.

Dr. Sage Claydon: With – one of the things and I know people tend to think, "Oh, well, we should always do what we've always done." But one of the things with our literature and our procedures is you know there's often codes that are created that don't actually apply to our literature of how people discuss them as we learn more about what we're doing and you know mesh certainly is cutting edge to be politically correct, so as we learn more about things that are going on specific to mesh, it would be very nice to have codes match the body of medical literature and the terms that physicians use.

Operator: Pardon the interruption, Dr. Claydon.

Dr. Sage Claydon: No problem.

Operator: The main presenter's line has disconnected.

Beth Fisher: Oh, gosh.

Operator: So I'm going to place everyone on music hold until we can re-establish that line.

Beth Fisher: OK.

Operator: Thank you.

Linda Holtzman: Then why are we having this code edition on note at 996.7, because it seems like 629 already gives you enough information about – you know, let's just assume that you know it's very clear at least to the physicians documenting it that they make a distinction between erosion and exposure. So I guess I don't understand Option 2.

Beth Fisher: Needing to use two codes, you mean?

Linda Holtzman: Yes. Meaning to use two codes, one saying it's a mechanic complication and the other one saying it's not.

Beth Fisher: OK. OK. Thank you. I don't know I can explain that, but I'll take the comment into consideration – a mismatch, so to speak.

                  I think we have Dr. Claydon back on the phone.

Dr. Sage Claydon: Yes. I'm back.

Beth Fisher: Sorry, we lost you somehow.

Dr. Sage Claydon: Yes. It maybe was a sign you know mid-sentence, I got disconnected.

Beth Fisher: I think we understood what you were saying and then we had a commenter here in the room indicated …

Dr. Sage Claydon: OK.

Beth Fisher: … that sometimes is a mismatch between how different specialties speak, terminology wise versus how the classification speak is. And I think we have to carefully look at those terms and how we add them to the classification or

whether we also have the option of waiting until we – putting it into ICD-10-CM which is a little bit more flexibility also. But is that what you were trying to say is this terminology mismatch sometimes?

Dr. Sage Claydon: Well, there's ultimate terminology mismatch between how things are written up for the coders and what the medical literature and the physicians call it.

So I put in pacemakers for the bladder and certainly, if a pacemaker – I would call that an erosion, not an exposure, if it's all of a sudden eroded through the multiple layers of tissue that I bury it under in the buttock.

So I don't know – as long as physicians are writing their notes, I don't think that's going to be such an issue. And you know with the vagina, it's become an issue in communication for us because there's really no other place – there's no other place that they put a polypropylene mesh directly under the skin. And so this is a specific – it's more specific to the mesh, even the general surgeons when they place hernia meshes, they're placed deep into the abdomen or deep into the abdominal wall, so it's not skin and mesh whereas for the vagina, it's skin and then mesh. And I think that's where it comes into play the difference and that's why ACOG asked for a different – you know, that distinction.

Beth Fisher: OK.

Dr. Sage Claydon: Does that make sense?

Beth Fisher: Well, I – yes, I understand what you're saying.

Dr. Sage Claydon: All right.

Beth Fisher: I can't agree or disagree or say we're going to do it one way or the other, but I can take all those comments into consideration, so I thank you for those.

Do we have any comments from the phone lines?

Operator: Yes, we do, from Susan Proctor. Your line is now open.

Beth Fisher: OK.

Susan Proctor: Hi. I have a clinical question and I apologize if it got discussed sometime
                   when …

Beth Fisher: Hello?

Susan Proctor: Hello.

Beth Fisher: Yes. Go ahead.

Susan Proctor: OK. I have a clinical question for the doctor.

Beth Fisher: OK.

Susan Proctor: Do these conditions exist at the same time in any patient?

Dr. Sage Claydon: Yes.

Beth Fisher: Oh, go ahead. You said that – can the two conditions exist at the same time?

Susan Proctor: Yes.

Dr. Sage Claydon: Yes, absolutely. They can be mutually exclusive or they can exist at the same
                   time. So, yes.

Susan Proctor: Thank you.

Beth Fisher: OK. Any other comments from our phone lines, operator?

Operator: There are no further questions at this time.

Beth Fisher: OK. If we have no more questions here in the room, I'm going to take all
                   these comments into consideration. It's a lot to think about. And so we may
                   see this again in March or – but we have a lot to look at.

                   OK. I'm going to turn the program back over – Dr. Claydon, thank you for
                   participating.

Dr. Sage Claydon: Oh, absolutely.

Beth Fisher: OK. We'll move on to our next topic.

Donna Pickett: OK. Our second topic for discussion this morning is on malnutrition which is on Page 18. Is Dr. Gordon Jensen on the line?

Dr. Gordon Jensen: Yes, I am.

Donna Pickett: Good morning, Dr. Jensen.

Dr. Gordon Jensen: Good morning.

Donna Pickett: The American Dietetic Association and the American Society for Parenteral and Enteral Nutrition are requesting new codes for malnutrition and the addition of several instructional notes to clarify the use of the codes.

The existing ICD-9-CM codes for malnutrition are outdated and do not reflect the current standard of care. In the interest of time, I won't read through all of the background information. You have it in front of you.

If there are any clinical questions about how the understanding of malnutrition has changed over the years and clearly not reflected in the current ICD-9-CM, I'd like to take questions from the floor.

And we also have representatives from the two organizations in the audience with us today as well as Dr. Jensen on the line so we have a good wealth of expertise with us on the phone and in the room.

Operator, are there any questions on the line?

Operator: If you have a question or a comment, please press star zero on your telephone keypad. There are no questions at this time.

Donna Pickett: OK. Dr. Gordon, is there some additional information that you'd like to bring to the discussion this morning before we move into discussion of the actual coding proposal?

Dr. Gordon Jensen: Sure. I just want to thank you for considering this proposal and you know I think our organizations feel that this is just fundamentally very important

and that the current approach to coding is just hopelessly outdated and this does not incorporate our current understanding of malnutrition or inflammatory response.

And we've had just immense requests throughout the United States from health practitioners, from coders, for us to address this issue. And so what you're looking at is a concerted collaborative effort between two leading nutritional organizations to do just that.

Donna Pickett: Thank you, Dr. Jensen. OK, we have a question from the floor. Please remember to state your name.

John Shaw: It's John Shaw from NextWave. It appears from the proposals that everything is moving in the direction of other specified or unspecified protein-calorie malnutrition and moving away from kwashiorkor and nutritional marasmus. Are those two terms, kwashiorkor and nutritional marasmus, still in use and still significant?

Donna Pickett: Dr. Jensen, would you like to address that question?

Dr. Gordon Jensen: Sure. They're in widespread use and misuse, and, of course, the proposal that is in front of you attempts to direct people to more appropriate use of codes ultimately with an eye towards – you know, as we move towards ICD-10 is moving towards disease and inflammation-based codes that's distinguished from pure starvation.

An important feature though of what we're proposing is that access to these more antiquated codes remains and will be in use and, of course, we're very interested to see ultimately how practitioners will continue to use them as we educate folks in regard to the new approach, but you know we're certainly not proposing to take them away immediately.

The reality remains that the – you know, whether it's marasmus, kwashiorkor, PCM, they promote tremendous confusion and misuse throughout the United States today you know people that are you know profoundly malnourished who were not recognized as such and vice versa.

John Shaw: I guess what would be helpful is to have a distinction of what is the correct use of the antiquated terms or is there a correct use of the antiquated terms?

Dr. Gordon Jensen: It's not clear that there is a correct use of the antiquated terms, and they have so much baggage associated with them that you know we have actually explored attempts to redefine them and all that did was create even greater confusion.

The reality remains that the new approach we're proposing easily accommodates all those historic constructs, but that said you know it certainly makes sense to have them remain available for the time being especially because it was with great interest to explore how they're being used and misused while we implement this new approach.

Donna Pickett: OK. We have another question from the floor.

Brigitte Kruder: Hi. Brigitte Kruder. My question actually is a coding question related to this proposal. Normally, when we have a chronic condition and we have something that's acute which in this case would be the malnutrition, we would sequence the acute condition first and I would look at the malnutrition, it's only directing attention towards the issue with the malnutrition that we would sequence that first as opposed to the chronic disease which would be the neoplasm or the chronic pancreatitis or whatever. We're not directing any attention at that. Why would we, particularly with the neoplasm, because in most instances, the guidelines will tell you that you sequence what you're treating if you're not treating the neoplasm.

So I guess the – I'm OK with the code first for the acute illness and all of that but with the chronic, I have a little bit of an issue.

Dr. David Berglund: This is Dr. David Berglund, NCHS. I wanted to respond to John Shaw on the appropriate use of these other two codes. I think most of us have probably seen pictures of starving children from various places and those would be – they would usually choose one either with marasmus or kwashiorkor. We do get that degree of malnutrition in United States on very rare occasions so it is possible that you could see those diagnoses, but it would be extremely unusual.

Dr. Gordon Jensen: Yes. That's correct. Jane, do you want to tackle the coding question?

Jane White: I'm not …

Donna Pickett: Jane …

Jane White: Oh, I'm sorry.

Donna Pickett: Introduce yourself.

Jane White: Jane White, American Dietetic Association. I guess I'm not sure that I really understand what you were saying about the code first and the chronic disease.

Donna Pickett: OK. Let me try to explain that. We do have some coding guidelines that indicate if you're only treating the – if your focus is treating the anemia and the person has a neoplasm that the anemia code would go – be sequenced first and that the neoplasm would be sequenced secondarily.

And so (Brigitte's) question is having instructions at the 262.13 and the 263.19 actually change how things have been coded over time.

Jane White: OK. In line of that, if you're treating malnutrition as the primary diagnosis then we would be certainly very pleased to have that coded first.

Donna Pickett: Not with the …

Jane White: Yes, no. I realize that. Yes, we didn't understand that. Thank you.

John Shaw: Hi, It's John Shaw again. I maybe a special case, but when I adopted my son from Haiti, he arrived with kwashiorkor in the United States.

Donna Pickett: Yes.

John Shaw: And a side comment I heard is that the appropriate – the more appropriate use of both of the terms tends to be in children rather than adult and it's inappropriately used in adults. So perhaps even more if those terms are typically inappropriate, some instructional notes that call them antiquated terms or more typical in underdeveloped countries or something to help push

the physicians that may still be using the old terms in the direction of the correct usage.

Donna Pickett: OK. Thank you, John.

Dr. Gordon Jensen: Yes. This is Dr. Jensen again. That's correct. And in more than 20 years of clinical practice as a physician/nutrition specialist, I've probably seen eight legitimate cases of kwashiorkor in the United States, often, that kind of scenario that was just described.

And I would add that we actually do have an ASPEN task force that's specifically looking at the diagnostic characteristics in regard to children that go with this as part of our educational efforts, so that sort of clarification will indeed be very important.

I think the key is that the proposed new approach really does accommodate children as well but we do think it's important that at least for the time being to have the option remain of using these antiquated diagnosis especially in the context of children.

Donna Pickett: OK, we have another question from the floor?

Linda Holtzman: Linda Holtzman. I guess these are like technical coding questions. First, I understand that malnutrition isn't spontaneous condition that's it's you know the patient is always going to have some kind of other process going on that's going to lead to malnutrition, but I'm wondering how would we code just the statement of severe malnutrition? I mean, in other words, is there a default here because I mean we have an acute and chronic and environmental, social and then an other?

Well, if I've you know got a chart that's several inches thick the patient's got a whole bunch of problems and then somebody writes "severe malnutrition" and doesn't attribute it to something, what would you like me to do with that?

And I guess part of that question also is are we going to need guidelines on how far a coder can go in associating it with other conditions there that are documented? You know, for example, if the patient comes in and I see that

the patient has severe malnutrition and the patient also has –- I don't know – ulcerative colitis and nobody actually documents that one is due to the other, can I go ahead and make that assumption? So that's you know something that needs to be looked at.

I guess the other question I have or just the point that I have is on 262.19, there's a code first note and it says, to code first a related disorder including frailty, 797 script, that's a symptom code. So we'd be looking at putting a symptom code as the principal diagnosis with a non-symptom code as a secondary. You'd have to put that.

Donna Pickett: OK. Linda, in response to a couple of your questions, the default for the severe malnutrition would be 263.9. That is in the alphabetic index piece of this proposal but the default for malnutrition was already 263.9 so that has been addressed in the alphabetic index changes that are in the handout.

Linda Holtzman: Yes.

Donna Pickett: And interestingly, when we were doing the proposal, we had discussions about frailty and where it should be or shouldn't be, so thank you for that question because I knew that would be a concern.

Another speaker at the floor here?

Jeanne Yoder: Jeanne Yoder, TRICARE Management Activity. There are a couple of other symptoms that may progress to malnutrition. The loss of the sense of smell and taste. Sometimes people becomes disinterested in eating and whatever and then there's also – and so that's 781.1, and the loss of appetite, the 783.0, would those also go under the – related to other disorders?

Donna Pickett: I would refer to ASPEN and ADA for that response.

Dr. Gordon Jensen: This is Dr. Jensen. That certainly sounds appropriate for us to consider. Those are symptoms that we use quite often to characterize malnutrition – so sure.

Donna Pickett: Thank you, Dr. Jensen. We have another question from the floor.

Nelly Leon-Chisen: Nelly Leon-Chisen, American Hospital Association. I see the index revisions where you do have malnutrition NOS and severe malnutrition going to the same default code. I think that's a problem because if it's severe where the physician has documented it as severe, to me that indicates a degree of you know more seriousness such as malnutrition NOS. And I understand you know saying that 261 may not necessarily be the appropriate default code but maybe we need to have a severe malnutrition with NOS.

Donna Pickett: OK.

Operator: Excuse me. We have a comment from the phones.

Donna Pickett: Thank you, Lisa.

Operator: You're welcome. From Susan Proctor. Your line is now open.

Susan Proctor: Thank you. I have an issue at the proposed 262.12. Do you have transplant complications listed …

Donna Pickett: Susan, we're not hearing you.

Susan Proctor: At 262.12 in the proposal, the transplant complication …

Donna Pickett: Go ahead.

Susan Proctor: ... is listed there. I sometimes see many things that are post-operative from GI surgery or other types of surgery or a procedure like transplants. And I would not call them acute illnesses. There may be a long-term type thing. I think maybe in this proposal there needs to be another – like 262.15 to include that kind of category.

Donna Pickett: OK. Susan, so are you saying that there are probably some conditions that really aren't acute, but they're really not chronic either, but you think those need to be represented as well?

Susan Proctor: I think they may well be chronic but they might not be specified as that. They're resulting from say, transplant complications or post-GI surgeries.

Especially people with the bypass surgery, we see a lot of them that have malnutrition coming in.

Donna Pickett: OK. Thank you, Susan. We'll take a look at that as well.

Susan Proctor: Thank you.

Donna Pickett: Are there any other questions on the call in?

Operator: Yes, we have a question from Dr. Linzer. Your line is now open.

Dr. Jeffrey Linzer: Thank you. I want to thank Dr. Jensen for making his comments and …

Donna Pickett: Dr. Linzer, we're having a hard time hearing you.

Dr. Jeffrey Linzer: I'd like to thank ASPEN and Dr. Jensen for the comments that they have made concerning these nutritional issues especially when it comes to children. I certainly agree that in the United States, some of these terms are becoming less common and weren't used but, of course, since we have to adhere to the (WHO) standard, we may not be able to completely remove these terms until that comes to us.

                   But as I've said at previous meetings, I think this is another example of where the educational process for medical students and residents really needs to kicked up a notch and that it's the training of the future clinicians in the proper terminology that is going to be a key to the appropriate use of these diagnosis codes.

Ainsley Malone: Can I make a comment to that? I'm Ainsley Malone with ASPEN and both ASPEN and ADA have task forces in place right now that have just started work on approach in medical education within medical schools. So this is one certain area where we hope to have an impact.

Dr. Jeffrey Linzer: And I congratulate you for that. I wish more societies would take such a proactive stand.

Donna Pickett: OK. Even though we sort of kind of worked our way through the proposal, I just want to go through it again very quickly. At 260, we would be deleting

the inclusion term there. It would remain in the index but would not be in the tabular. The inclusion terms at 261, again, would remain in the index but would not appear in the tabular and the severe malnutrition, thank you for the comments on that and we'll look at addressing how best to index that particular term.

The expansion at 262, we're modifying the category title and now, it would be other specified severe protein-calorie malnutrition. We're creating a new subcategory 262.1 and new fifth digits under 262.1 – 262.11, severe malnutrition and acute injury, at the point 12, in acute illness, at the point 13, in chronic illness and at the point 14, in environmental and social circumstances. At the 262.19, what did not end up appearing in the final version of this proposal, it should read severe malnutrition and other disorders.

And at the 262.13, frailty is listed there, and I know we've had a discussion about placement of frailty and the fact it is a symptom but it would be – it should have been deleted from the list of conditions at the 262.13 but we will take into consideration what to do with the 262.19.

And then you had the other modifications as shown and again, the alphabetic index changes, which is to give you an idea of how the index entries would change for the revisions to the 260, 261 and the expansion of the 262.

Are there any other questions from the floor on this proposal? And we have one person coming to the microphone.

Linda Holtzman: Linda Holtzman. Mine's actually a clinical question. Maybe you can address it? I just want to get a little bit more background information on what exactly you mean by severe malnutrition and acute injury and severe malnutrition and acute illness. I have to say I don't usually see malnutrition associated with an acute admission. At least from my own experience which may not be representative, it's typically you know a chronic situation. So if you could give some examples of that and how that works?

Dr. Gordon Jensen: Sure. This is Dr. Jensen. What you're speaking to is the challenge of discerning when malnutrition actually occurs or is present. And some of our

colleagues attempt to refer to you know massive acute inflammatory response and in trauma or surgery or illnesses, acute metabolic dysregulation which is a whole another mouthful and you know would be whole another confusing diagnosis to bring to bear.

But the reality, of course, is that the acute inflammatory response triggers a host of events and that includes acutely you know marked erosion of muscle mass and a host of other metabolic changes.

And so rather than attempt to you know draw a line the sand and say, "Well you know you're malnourished on you know day five as opposed to day two." Much simpler approach is to identify anybody with a very acute significant injury inflammatory illness response as being at great risk for an adverse outcome.

So that is the approach and, of course, you can see what we've done here is make this really all ultimately etiology-based. And the bottom line being that anybody with a very significant acute injury or illness warrants some very close evaluation and follow-up rather than waiting for them to become you know severely malnourished in the classic sense. And so that's the simplicity of this present approach.

Donna Pickett: Thank you, Dr. Jensen. In the interest of time, I remember I said I was going to be the timekeeper here; I will invite everyone to provide your comments in writing on this. I see that there are probably some other people that have thoughts on this issue, and welcome and encourage you to send us your comments on this.

With that, I'm going to close the discussion on the malnutrition proposal and turn the podium back over to Beth Fisher who will handle the next couple of topics. Thank you Dr. Jensen.

Dr. Gordon Jensen: Thank you.

Beth Fisher: OK. Then we'll turn to Page 25, and I'd like ask if Dr. Jon Hathaway is on the phone? If you could unmute your phone and hopefully you've dialed in.

Operator: I do not see his line at this time, however, if you are on the line, please press star and the number one on your telephone keypad.

Beth Fisher: OK. Operator, I think he may have been dialing in on a separate speaker phone. Dr. Jon Hathaway, are you there? If not …

Operator: Your line is now open, Mr. – Dr. Hathaway.

Dr. Jon Hathaway: OK. I'm here.

Beth Fisher: OK. Hello, Dr. Hathaway. Thank you for joining us. We're going to cover the topic of the – we titled it elective C-section prior to 39 weeks. I don't know if that quite – it describes the scenario but this topic does come from – again, from the American College of OB-GYN, the ACOG folks that we are in touched with quite often.

And Dr. Hathaway, maybe you could give a description of this for the audience. I have given a slight – a small description in our topic packet here of the – if I understand it correctly, it's primarily – well, it's brought up as a quality issue that you're trying to better track repeat C-sections that may occur prior to 39 weeks so maybe you could give us a little bit more background on that.

Dr. Jon Hathaway: Right. Correct. Currently, there is no ICD-9 code that would distinguish between an elective repeat C-section between 37 and 39 weeks and the repeat C-section that occurs during that same time for a patient that's in labor and there have been several articles recently in New England Journal of Medicine as well as the American Journal of OB-GYN that have pointed out that performing an elective repeat C-section prior to 39 weeks increases the neonatal morbidity.

So a lot of hospitals as well as government agencies and insurance companies are tracking which physicians are performing these procedures as well hospitals are granting and denying privileges based upon whether a physician is performing this procedure prior to 39 weeks electively.

So recently, our hospital in Indiana University was audited and we found that we had a significant number of these elective C-sections between 37 and 39 weeks. But then when – as physicians, we went back to the charts, we found that most of these patients were actually in labor and there is no code to say we performed the C-section because she was actually in labor.

Beth Fisher: OK. So we looked at the codes and for that time period that you're looking at, it appeared that we've got codes for early labor specifically – I don't have my codebook in front of me. I'm at a loss, but …

Dr. Jon Hathaway: I have one in front of me if you'd like me to …

Beth Fisher: Early labor before – you know, before 37 weeks and I think even we've got the false labor code also where you've got labor without delivery but we've got this small window of between 37 and 39 weeks, that's really not covered.

So we were trying to come up with a way to show this with a new code which we were proposing at 644.3 with just the zero and one and the title is kind of difficult to come up with too. It's – we can't really say early labor because we've already got that in another code so we said late preterm onset of labor with delivery with the inclusion term late preterm onset of labor after 37 completed weeks of gestation but before 39 weeks with delivery and then the same with onset of labor after 37 weeks but before 39 weeks with delivery.

Doesn't specifically say it for C-section because we were, I guess, trying to not combine those two axes together. So I'm going to go ahead and take comments from the floor because I see that Sue is at the microphone.

Sue Bowman: Sue Bowman, AHIMA. And Nelly and I are just wondering if perhaps we haven't had enough coffee this morning or else we've had too much – one of the two. But late preterm to me sounds like an oxymoron.

In the category of early or threatened labor, so I admit to hopeless confusion …

Beth Fisher: And so the question is where to put it also, you're right in this chapter. So, Dr. Hathaway, what – I mean do you have any suggestions for how to title this, because it is difficult?

Dr. Jon Hathaway: It is difficult because there's also late preterm labor in the literature which is defined as 34 to 37 weeks by some people and not by everybody. There's no standard nomenclature for these definitions and when you look at the literature where they talked about this 37 to some 39-week delivery and where they were tracking things, they didn't come up with a good name either. They just called it prior to 39 weeks and after 37 weeks so …

Beth Fisher: So, OK. Aside from the title, maybe where we have to come up with a good title. Is this – I guess I ask the audience here in the room. Is this – do you see this happening often? And maybe Dr. Hathaway could – you said that you looked at this in your hospital? How often where you say this happens of the early C-sections that you saw? What percentage maybe of them are in this scenario you're describing?

Dr. Jon Hathaway: Well, when you look – when people have looked nationally, so I'm specifically thinking of The New England Journal of Medicine article that came out in 2009. They found that over half of repeat C-sections were performed between 37 and 39 weeks. So this is a significant number.

Beth Fisher: And then of those, what percentage do you know are the ones that are in labor versus they just – they did early – I don't know – it's just kind of hard to come up with the percentage of how often this happens, I guess.

Dr. Jon Hathaway: So they found also – it's a similar number so about 50 percent of them were had an indication and 50 percent didn't but part of the problem with that is how do you track that. So the Netherlands did a seven-year study where they were able to show that it was about half of these C-sections performed between 37 and 39 weeks were done electively without labor – but that's in the Netherlands, that's not in the United States so it's difficult to track because we have no way of coding it.

Beth Fisher: OK. So title is a challenge. Do we have any comments from other phone participation – participants, operator?

Operator: If you would like to ask a question on this topic or any other, please press star and the number one on your telephone keypad.

We have a question or comment from Dr. Linzer. Your line is now open.

Dr. Jeffrey Linzer: Thank you. I appreciate the dilemma of trying to track this issue and I'm wondering that instead of using the term late preterm labor or something similar …

Beth Fisher: OK.

Dr. Jeffrey Linzer: … if it would just be easier to indicate that the number of weeks that the delivery occurred at starting at, say, 30 or 31 weeks up towards term or 38 weeks and use that to …

Donna Pickett: Dr. Linzer, it's hard to hear you again. I don't know if we have a bad connection. Could …

Dr. Jeffrey Linzer: I'll try it again. I'm just wondering instead of using the term late preterm labor, if it would be better to indicate by number of weeks when the delivery occurred and whether it was with active or without active labor.

Beth Fisher: OK. We'll have to look at that a little bit. It kind of alludes to a separate proposal for the weeks gestation and I see the point but OK, we can take that comment.

OK. Do we have any other comments on the phone, operator?

Operator: There are no further comments at this time.

Beth Fisher: OK. We have one here in the room.

Kim Seery: Hi. This is Kim Seery. We were just discussing possibly the addition of a V code for a patient in labor versus not in labor and still continuing to use the 654.2 as an option.

Beth Fisher: V code, you said?

Kim Seery: Yes.

Beth Fisher: Yes. OK.

Dr. Jon Hathaway: My understanding of a V code is that it's for a potential event, not an actual condition at the time and …

Beth Fisher: Well, not necessarily, no.

Dr. Jon Hathaway: OK. I would propose that a woman who's in labor says she's really having an issue at that time.

Beth Fisher: Yes. I'm not sure a V code would do it either but we – yes, OK. OK. I hope there are no further comments. Thank you, Dr. Hathaway, for helping us with the discussion, anyway. We'll have to look at this a little bit further.

Dr. – sorry, I have one more comment in the room here.

Kathy Rivera: Kathy Rivera. I was just curious as to why there is not the fifth digit of two.

Beth Fisher: Yes, yes. That's – I was curious – a postpartum condition with delivery or …

Kathy Rivera: It's delivered with a postpartum …

Beth Fisher: Yes.

Kathy Rivera: ... problem.

Beth Fisher: Yes, you're right. I saw that and I thought maybe we should add that too. Actually, I was looking at our tips and we don't have any – I mean we can add it.

Kathy Rivera: OK.

Beth Fisher: But it looks like we don't have a lot of combinations. We're too – it's either like zero through four or 01 or 013 but …

Kathy Rivera: Zero-one-three, yes.

Beth Fisher: ... you're right.

Kathy Rivera: Yes.

Beth Fisher: Yes, I looked into. It looks like it could apply as well so ...

Kathy Rivera: Yes, yes.

Dr. Jon Hathaway: What would be the postpartum complication now?

Beth Fisher: Well, it would be showing that they delivered, but that they have – but yes
                     you know this wouldn't be a postpartum complication though.

Dr. Jon Hathaway: Because most of the time, the mother does fine if the …

Beth Fisher: Yes.

Dr. Jon Hathaway: ... resulting neonate that has two times the morbidity of other you know
                     neonate.

Beth Fisher: OK. All right. And then just as a coding question, then you would use the
                     repeat C-section with these codes?

Kathy Rivera: Yes. I think you've said …

Dr. Jon Hathaway: For the repeat C-section?

Beth Fisher: You'd still want to use the repeat C-section along with this code.

Dr. Jon Hathaway: Correct.

Kathy Rivera: OK.

Beth Fisher: OK. Oops, we have another comment, I think, coming to the microphone.

Wendy Droppleman: I'm Wendy Dropelman. I'm wondering if this is really an issue for the
                     pediatric patient. Why we're not looking in a neonatal code instead of a labor
                     code?

Dr. Jon Hathaway: I'll defer to the pediatricians for that. The reason the obstetricians are looking for a code is mainly because we're being held accountable for our timing of elective repeat C-section and sometimes our hand is forced by the onset of labor and there's no way of documenting that in the – with a simple ICD-9 code.

Beth Fisher: Yes. But the request here – the request came from the obstetricians so we hadn't received a request from the pediatricians but …

Dr. Jon Hathaway: I'm sure there are plenty of codes in the neonatal section about respiratory distress and what other actual complication occurs for the neonate.

Beth Fisher: OK. All right. No more comments in the room?

Operator, are there any more comments on the phone?

Operator: Yes. We have Dr. Linzer. Your line is now open.

Beth Fisher: Dr. Linzer, hello.

Dr. Jeffrey Linzer: Hi. Well, speaking for the Academy of Pediatrics, I think that we have a good code set to indicate the gestational age of the child and I certainly agree with my colleagues from ACOG that it's important to be able to track from their end when the C-section is being done and under what conditions. He's absolutely correct; it does contribute to the number of children who end up in the NICU and who end up with a problem.

So, certainly, I agree that while there are specific codes for newborn children prematurity, there needs to be something to help the obstetricians track this very important issue.

Beth Fisher: OK. Thank you. All right. No more comments?

Operator: There are no further questions on the phone.

Beth Fisher: OK. Dr. Hathaway, are you able to stay on the phone for a couple of more – two more of the ACOG topics?

Dr. Jon Hathaway: Sure.

Beth Fisher: In case we have any questions for medical-related – we'll move on to the next page of our topic. It's Page 26 and that is a request for a code for personal history of gestational diabetes that ACOG requested and we're proposing to add this to the V12.2 with the V12.21, personal history of gestational diabetes and then V12.29 for personal history of other endocrine, metabolic immunity disorder.

Dr. Hathaway, did you have any more comments on the importance of tracking this that we didn't really have any other you know medical information from your group?

Dr. Jon Hathaway: I think – and I'll defer to Donna if she's out there to chime in.

Beth Fisher: Donna Tyler is from ACOG here in the audience, yes.

Dr. Jon Hathaway: So, my understanding when we discussed this at our meeting was that when a patient comes in with a gestational diabetes in her prior pregnancy, we need to be able to screen them for diabetes in the current pregnancies, since over half of them will develop type 2 diabetes within the next four years of having a pregnancy with diabetes.

However, there is no – again, no code that allows us to tell the insurance company we're doing this screening test prior to 28 weeks because of her prior history of gestational diabetes and there's no way – and so what was happening was codes are being denied because we were doing two diabetes tests during one pregnancy – one early and then one at the appropriate time of 28.

Beth Fisher: OK. So basically, because of the potential for developing type 2 from past history. OK.

Dr. Jon Hathaway: Correct.

Beth Fisher: All right.

Dr. Jon Hathaway: And we would manage those patients differently earlier in their pregnancy and not wait until 28 weeks to initiate treatment for their non-gestational diabetes.

Beth Fisher: OK. All right. Are there any comments in the room on this proposal? Are there any comments on the telephone, operator?

Operator: Not at this time.

Beth Fisher: OK.

Dr. Jon Hathaway: Well, that was much easier.

Beth Fisher: Yes. This next one may be a little more – maybe not, I don't know if – I shouldn't say that.

Page 27 is the request for two new codes that are somewhat related, I think, the way it was presented to us from the American College of OB/GYN, and that is a code for a history of ectopic pregnancy and then a code for, basically, wanting to show a reason or an encounter for a patient having usually an ultrasound, I guess, to confirm fetal viability early on in pregnancy. And that sometimes early on a pregnancy, a heartbeat can't be detected but the patient has a history of an ectopic pregnancy so they need to have an ultrasound and so we are wanting to propose two new codes – one, for the history of the ectopic pregnancy at V23.42 and then a code for a pregnancy with inconclusive fetal viability or an inclusion term of encounter to determine fetal viability in pregnancy.

And Dr. Hathaway, did you have anything to add to this background?

Dr. Jon Hathaway: No, I think that's pretty consistent. You sometimes have patients show up with lower quadrant pain and you wonder if this is an ectopic pregnancy that's rupturing, a follicular cyst that's rupturing, corpus luteum cyst or just that she ate something bad last night for dinner. So – but if someone has a history of an ectopic, they're at much higher risk of having a second ectopic pregnancy. And so in order to confirm that, the best way is by ultrasound and we already

do entirely too many ultrasounds during pregnancy and so this would be one way of saying this is a necessary one.

Beth Fisher: Oh, OK. OK. Comments from the floor?

Sue Bowman: Sue Bowman. I'm just wondering if the proposed V23.43 is in the right place and you know we're talking about that…

Beth Fisher: Yes.

Sue Bowman: … but this time, I think it was poor obstetric history, meaning a previous pregnancy.

Dr. Jon Hathaway: Right. This would be a previous ectopic pregnancy.

Sue Bowman: No, the other code.

Dr. Jon Hathaway: Oh, I'm sorry.

Beth Fisher: Pregnancy with inconclusive fetal viability.

Sue Bowman: I mean, that's like now, right?

Beth Fisher: Yes. So we're …

Dr. Jon Hathaway: OK.

Beth Fisher: Well, I guess, we're putting it within the supervision of a high-risk pregnancy within that.

Sue Bowman: But the way – it's in the V23.4, which is pregnancy with other poor obstetric history.

Beth Fisher: OK. So location, yes. Location, location, location. But did you have any comments about the need for the code itself or one way or the other? OK, all right.

Any other comments here in the room? David now has the topic packet on the screen for those who may wish to follow there.

Do we have any telephone comments, operator?

Operator: If you have a comment or a question, please press star and the number one on your telephone keypad. There are no questions or comments at this time.

Beth Fisher: OK. Thank you.

OK. Dr. Hathaway, that's all of the topics for now. I mean, thank you for joining us.

Dr. Jon Hathaway: Oh, thank you for having me.

Beth Fisher: And we'll move on to a different topic altogether, which come from the American Academy of Neurology, so I'd like to ask if Dr. John Hart – we have presenters in the room. He's – comes to us from the American Academy of Neurology to present the corticobasal degeneration. So, I guess, I should ask Austin if you could load that presentation that I gave you earlier?

Austin: Yes.

Beth Fisher: Very good.

We have two topics to cover for Dr. Hart. The first one is on Page 36, corticobasal degeneration. We'll discuss that one and when he's – we're finished with comments on that, we'll move to a second topic on visual agnosia. So I'm going to turn it over to Dr. Hart.

Dr. John Hart: Thank you, Beth. Thank you, everyone.

As we've been exploring and better clarifying both topologically and clinically, the dementias that occur in terms of both looking at the clinical manifestations and the pathology, there's been a growing number of well-defined illnesses that unfortunately have not made their way into the coding system, one of which is corticobasal degeneration.

It's a neurodegenerative disease and it's a combination both a movement disorder disease and a cognitive disorder. The cognitive disorder looks

someone like frontotemporal dementia. One of the most important aspects is there's usually a speech problem that's more relevant and present in these patients than even in the FTD, frontotemporal patients or Alzheimer's patients and they also have a movement disorder. So you'll see the patient and they'll have mostly a loss of executive functions, which are things like organization and planning and being able to focus attention. They have visual-spatial problems and will get lost rather easily and then the language impairment as we noted.

The movement disorder is also unusual and atypical in that it's mostly asymmetric and the hallmark feature usually is apraxia, which is when you tell someone to do something and their motor system, they can't perform and do it. They can do it without being told, but they can't do it when you give them instructions and that's because their language system and their motor system are somewhat disconnected.

Otherwise, the patients can have dystonia, which is more of a writhing and uncontrolled movement of the arm and now, and alien limb syndrome where they don't realize that that is their arm that's present, jerking myoclonus, a loss of sensation or rigidity syndrome that goes with this – almost – always starting asymmetrically and then progressing as the disease progresses.

Clinically and pathologically, it's different from the other diseases that we have. We've been adding diseases for frontotemporal dementia over the last several years to clarify them. This disease both had a movement component, a cognitive component and it's typically classified and pathologically as having cal proteins which is the way we're starting to classify dementias and also, in the future, how the play in treatment is going in terms of looking at these disorders so it is unique in those aspects compared to the other codes we have.

And since ICD-9 does not include corticobasal degeneration, we posed to add the new code 331.6 for corticobasal degeneration and we've given an alternate, if possible, G31.3 or in 31.85 as another possibility although it sits well with the codes we've been adding in terms of 331.6. And I'll stop on that one and take any questions.

Beth Fisher: Yes. I guess, I should clarify. He was referring to ICD-10-CM because I did give a reference or recommendation of location that's the G31 code. Are there any questions for Dr. Hart on the condition itself or clinical – other clinical questions here in the room?

And if not, operator, are there any questions on the phone line for Dr. Hart?

Operator: Please press star and the number one on your telephone keypad. There are no questions at this time.

Beth Fisher: OK. Any comments about the code proposal? OK. Then why don't I let you continue on with Page 77.

This is actually an ICD-10-CM request. As we have. Dr. John Hart here right now, we thought we'll just have him cover them both and this is on visual agnosia and related conditions. So why don't you go ahead?

Dr. John Hart: Thank you.

These three conditions that we're presenting together, clinically, coexist. And, I guess, one of the major reasons that we felt that these were important is because of the disability and the different type of disability that the patients have when they acquired these disorders. They're also very hard to explain to other individuals and I've had patients who have visual agnosia who very clearly wanted to be declared because of their problems as blind.

And what the disorder is, is that while someone could see perfectly fine and their eyesight is perfectly OK, they do not have the ability to recognize objects in front of them in the world. And the reason this is, is while their perceptual system of seeing all the colors, lights and edges that you will detect, the part of their brain that puts them together to make objects is not functional. And as a result of that, while they do not bump into things as they walk around and see them, they can't identify what they are, they can't pick them up properly and they are quite often confused of what is in front of them in the world around them.

It is not due to any other problem in terms of vision or language or general mental decline. It is simply a fact of not putting together information that comes into the brain.

Prosopagnosia is a subtype of visual agnosia and it is one of the most unusual sort of things is very disabling. And also, people cannot recognize faces, and these are faces of familiar people that they see all the time. So the way people get around this and this is actually more common, we think than we can diagnose previously is they watch people's gait and they smell their perfume, and they look at other features of them to identify family members. And I've had patients clearly who will look if I keep everyone still in the room and put their wife, their nurse and two of the medical students in front of them and say, "Pick out who's your wife," that they cannot do it all. You let the person move and they figure out from the gait pattern.

So this inability to recognize familiar faces is a subtype, in some regards, of the visual agnosia. We can't recognize objects.

And the last one, simultagnosia that we like to present occurs more commonly in both people with degenerative disorders, as well as focal brain disorders and that is that they can only attend to one object at a time in front of them. So what will happen is, is that they will be confused easily when there are multiple things in their visual field that they have to choose from and use – very difficult.

In terms of driving, they really can't drive at all because they can't pick out the things in front of them unless they focus on one thing at a time. So at the end, what they have to do and we have to do with them to help them out is find ways that they can navigate to their world by reducing the information in front of them. So all three of these disorders are higher order beyond vision. Vision is normal in all of these folks. It's the ability to take the visual information and put it together to be meaningful information at the end.

Visual agnosia and prosopagnosia are currently coded in ICD-10-CM as a carryover from ICD-9-CM under H53.16. It's psychophysical visual disturbances.

In actuality, while psychophysical is a descriptor of this, this really is better described as what's called a symbolic dysfunction, meaning you could see all the information. You just can't integrate it into the symbols they are in the real world and so we think this better captures what's actually going on with these patients.

And the AM promotes deleting the above terms from H53.16 and adding the new code, R48.3 and putting these three things together – visual agnosia, prosopagnosia and simultagnosia since they are all basically a subtype and grouped together with the visual agnosias where one is multiple objects, the other is faces and the other is objects in general.

Beth Fisher: OK. Do we have any questions for Dr. Hart in the room on this proposal? When he spoke about it being a carryover from ICD-9-CM, we did index these terms. I remember specifically, prosopagnosia being indexed since when we carried it over to 10-CM, we indexed it to the similar code in 10-CM but, as he is pointing out, it's better placed.

The R48 is in the symptoms chapter. I guess, I have one little question. And so how do you distinguish this from, say, an Alzheimer's patient who just doesn't remember their family members or something?

Dr. John Hart: Right. And the way you do that is if you have a – these are not memory problems. So if you have a patient who has Alzheimer's disease or other disorders causing an inability to, say, recognize someone's name, so you put two people in front of them and say which one – is this your wife, is this your wife? Or is that Marilyn or is that Marilyn? You take the memory component out and they speak and say, "Yes." You know, hello? They immediately now will remember who they are.

These folks don't recognize at all who these individuals are. No matter how much information you give them and how much information in queues visually, they have no idea who the people are in front of them. They know there are people there in prosopagnosia but they do not put together their faces in a way to recognize them. So we distinguish that memory problem by just giving them a recognition memory and say, "Is that your wife?" And she's

standing there and doing things, and Alzheimer's patients will immediately get that.

At very late stages, in any of the degenerative disorders though, these symptoms can appear. Usually though, they are in a host of a multitude of other things. These symptoms do appear after neoplasm, after stroke, after encephalitis and other illnesses as isolated phenomena that are permanent and non-changing.

Beth Fisher: Right. OK. Any comments on the coding proposal here in the room? OK. Then I'll ask the operator to open up the phone lines if there are any comments or questions.

Operator: Certainly, if you have a comment or a question, please press star and the number one on your telephone keypad. We do have the line of Linda Small. Your line is now open.

Beth Fisher: Go ahead.

Sharon Whitmore: Yes, hi. This is Sharon Whitmore. Actually, there's a whole group of us here. But we were just wondering, with these conditions, when do they occur and what causes them? Are they caused from birth or is it a degenerative type thing? Could you explain a little more of that for us please?

Dr. John Hart: Typically, they recur in focal brain injury or illness. So when you acquire the patients that I've taken care of with visual, prosop- and simultagnosia have had strokes, have had a heart attack and hypoxia or ischemia, have had status asthmaticus and also had a secondary problem with apoxia and have had encephalitis. So after brain damage to these regions that encode for this information, you're left with an inability to identify objects or people or multiple objects at once.

There are other conditions although much less common that I just mentioned earlier that are the late stages of degenerative diseases when these symptoms can appear, but they appear with a multitude of other symptoms. What we're typically talking about here are the focal sort of stable conditions that can

cause these symptoms. Again, if one wanted to and looked at late-stage Alzheimer's, Lewy body and other diseases, you may well see these also.

Beth Fisher: Did you have any further question? Treatment or any treatment? Probably not.

Dr. John Hart: You know, isolated conditions. It depends on the etiology. So in herpes simplex encephalitis, we treat patients with acyclovir and other medications for that.

In stroke, I must admit as chronic condition, it's very difficult to treat them. And if they appear in any of these other related degenerative conditions, the treatment would be for the primary disorder.

Beth Fisher: So OK.

Lisa Taylor: This is Lisa Taylor. If these symptoms have an etiology, maybe we should consider instructional notes for delayed effect or code first the underlying …

Beth Fisher: Yes, I'm thinking the same. I'll probably have to look at that, yes, this coding. OK.

OK. If there are no further comments, I don't think there are any.

Thank you, Dr. Hart. And John – I'm turning it over to David now.

Dr. David Berglund: All right. Next, we're going to be having a presentation by Mikhail Menis, presuming that he is on the phone.

Mikhail Menis, if you're on the phone, can you please speak up?

Operator, can you check the phone lines for Mikhail Menis?

Mikhail Menis: Sure. I'm on the phone. Thank you, Dr. Berglund.

Dr. David Berglund: All right. And Austin, can we have – yes, we do have Dr. Menis' presentation. All right.

And Dr. Mikhail Menis is with the FDA and he is going to talk with us about some issues related to transfusions. And Dr. Menis, I'll turn it over to you?

Mikhail Menis: Sure. Thank you so much, Dr. Berglund.

And my name is Mikhail Menis and I'm an epidemiologist from the Analytic Epidemiology Branch within CBER/OBE. And the topic of my presentation is to propose changes to the ICM-9 coding for transfusion associated adverse events.

The CBER's mission is to ensure safety and efficacy of biological products, including blood and blood products. And under the Food and Drug …

Dr. David Berglund: Hold a second. We're having a little trouble hearing you. Can you …

Mikhail Menis: Sure.

Dr. David Berglund: ... either speak a little louder or, Austin, can you turn the volume up a little for us?

Austin: Yes.

Dr. David Berglund: And just maybe slow down a little for people to – sorry, Mikhail.

Mikhail Menis: Sure.

Dr. David Berglund: And right now, we are showing your first slide. Let me know when you want to advance, and I'll turn it back to you now.

Mikhail Menis: Sure. So under the Food and Drug Administration Amendments Act, FDAAA, of 2007, in this response we're also conducting U.S. population-based active surveillance on medical product safety.

Now, currently raising medical databases from the Centers for Medicare and Medicaid Services health maintenance organizations, as well as private health insurance to conduct active surveillance, as well as identifying uncharacterized adverse events associated with transfusion of blood and blood products.

So the CBER has submitted a proposal – that's the next slide, requesting the addition of new specific ICD-9-CM diagnosis codes for transfusion associated adverse reactions, including anaphylactic reaction due to transfusion, other serum reaction due to transfusion and infection following transfusion, infusion or injection of blood and blood products.

So you introduced those new codes that will improve the precision of recording for transfusion associated adverse reactions and enhance this ability to conduct active surveillance of transfusion safety.

Overall, these codes will help in development of better transfusion-related reduction and prevention strategies. And so the next slide five is a specific topic for anaphylactic transfusion reactions. And those are the acute reactions due to antigen antibody interaction between donors and recipients of blood easily occurring within one to 45 minutes of transfusion with a major clinical manifestation of pruritic urticarial lesions, dyspnea, wheezing, intractable hypertension and shock.

Major mechanisms include Ig antibody interaction with proteins resulting in activation of mast cells and basophils and the release of anaphylotoxins. There's another mechanism of interaction of performed IgA antibodies in the IgA-deficient recipients with blood of non-diffusion donors causing the clinical manifestations.

And so the current coding, that's the next page, the current ICD-9-CM coding of 999.4 anaphylactic shock due to serum is not transfusion-specific and so we proposed a specific code, anaphylactic reaction due to administration of blood and blood products additionally because anaphylactic reactions can happen with vaccination, and it is important to monitor. There was – we also proposed an additional code, anaphylactic reaction due to vaccination.

And perhaps Dr. Berglund wanted to go further …

Dr. David Berglund: Certainly, are you – I'll talk a little more about the codes here …

Mikhail Menis: Yes.

Dr. David Berglund: ... for dyspnea.

Mikhail Menis: Yes.

Dr. David Berglund: And for those who are calling, one with the handout, we do have a number of things on those. I'm not going to try to go through all of them. What we've been discussing at this moment has been on page 12 of your handouts actually if you have a handout, and we would propose to change 999.4 instead of anaphylactic shock due to serum. We would change this to anaphylactic reaction due to serum. Anaphylactic reaction is included here but it's actually a broader term and we would propose to make a similar change for a number of other codes which are labeled anaphylactic shock but actually include anaphylactic reaction.

Now, in addition to that change, we would add new codes – 999.41 for the anaphylactic reaction due to administration of blood and blood products, 4.2 for anaphylactic reaction due to vaccination and 4.9 for anaphylactic reaction due to other serum. So those are the changes we've just been discussing.

There are some other serum reactions, and I'll turn it back to Dr. Menis to talk about those next.

Mikhail Menis: Sure. The next topic is the other serum reactions due to transfusion. And serum sickness is a hypersensitivity reaction resulting from a production of antibodies again for impotence and resulting in circulating immune complex deposition and causing fever, continuous eruptions and lymphadenopathy usually within one to two weeks after exposure. But in pre-sensitized individuals, it could happen within two to four days. The mechanism, as I mentioned, is a reaction between the recipient's antibodies and serum protein resulting in immune complex formation and deposition with the clinical manifestations.

And currently, the current ICD-9-CM coding of 999.5, other serum reaction, is not transfusion-specific so I proposed to add a new transfusion-specific code to read other serum reaction due to administration of blood and blood products.

And additionally, because other serum reaction can happen with vaccination, we propose that an additional coding of – to read other serum reactions due to vaccinations. So you know those are the two specific codings and perhaps, Dr. Berglund …

Dr. David Berglund: Certainly, I'll talk a little more about these.

On the codes we have proposed there then, we have 999.5, we would propose these two codes – 999.51. And actually, on the pages we have on page 12 of your handout, we have slightly different numbering of the codes. We have a 999.51 for other serum reaction due to administration of blood and blood products and 999.52 for other serum reaction due to vaccination with also 8.59 for other serum reaction.

Now, those are the codes that we are proposing to add. Sorry, what was – if you have questions, I'll ask you to come to the mic. There are you know the difference between what we have on the handout that we have on our Web site, in this slide, I'm afraid on the numbering, so it's point 51, point 52 and point 59 instead of point 50.

There are a number of other proposed changes here which primarily are changes in code title. For example, at 995.0, we have other anaphylactic shocks. That title will be changed to other anaphylactic reaction. This is on page 11 of the handout that we have.

And we would also change it, 995.6. We have anaphylactic shock due to adverse food reaction. We would change that to anaphylactic reaction due to food, and we would give specific code titles. For example, 995.60 would then become anaphylactic reaction due to unspecified food and each of these titles would be spelled out also as part of the proposal.

We would also change exclude notes in a large number of places to match with the new codes and code ranges that we are creating here in this proposal. And there are also a number of index modifications for codes, primarily, emphasizing use of the term "anaphylactic reaction" although anaphylactic shock could be continued to be indexed as it was before too.

Any comments or questions on this proposal on changes to the anaphylactic codes? I'd invite people to come to the microphone and invite comments.

Nelly Leon-Chisen: Nelly Leon-Chisen, American Hospital Association. We just – coming this October, we have a bunch of new codes in the 999.6 section where there are different types of incompatibility reaction due to transfusion of blood and blood products. We probably need to sort of have some sort of exclusion notes so that it doesn't get confused with the 999.5 one where – of the serum reaction due to administration of blood and blood products. You know, if someone could see the reaction and if the blood you know there needs to be some little bit of distinction between these codes.

Dr. David Berglund: So concerned about potential for overlap with the others and need for an exclude note, we'll be looking at that further again too.

Other comments? Do we have anyone else? No one else commenting right now here.

Operator, could you please check the phone lines for any comments?

Operator: Yes. If you have a question or a comment, please press star-1 on your telephone keypad. We have Dr. Linzer on the phone. Your line is now open.

Dr. Jeffrey Linzer: Thank you very much. I agree that the main change is going to be very important here since anaphylactic shock is, in and of itself, a bit archaic. And if you look at the papers that have recently been published in the allergy literature, the term "reaction" is more appropriate and broader.

My main concern is the sub-index term. While in the index you have proposed to put anaphylactoid reaction to the appropriate allergic reaction or anaphylactic reaction, I would suggest that you would add them as an inclusion term in the tab as well. Some clinicians feel that anaphylactoid is the term to use for non-IgE-mediated reaction but the reality is, clinically, you cannot differentiate them and they are treated exactly the same.

So certainly, a coder would have no way of knowing the difference between an anaphylactic and an anaphylactoid reaction if that tells the physician, documented it. And I think, while it's in the index, it may be of assistance to have it in the tab as well.

Dr. David Berglund: OK. So we did have that in the index but we can certainly consider putting that in. Now, would you suggest that with all the anaphylactic reaction codes that all of them at 995 and 999, Dr. Linzer?

Dr. Jeffrey Linzer: Yes, I would because …

Dr. David Berglund: You would, OK.

Dr. Jeffrey Linzer: ... they're going to be – that's diffusely used.

Dr. David Berglund: OK. So that's certainly something we can potentially look at adding. Well, I'm glad to hear from you, Dr. Linzer. We miss having you here, of course.

Dr. Jeffrey Linzer: Sorry, there wasn't room for one more.

Dr. David Berglund: All right. Any other comments on the phone lines?

Operator: There are no further questions or comments at this time.

Dr. David Berglund: OK. We have no further comments or questions here. We will next be moving on – moving forward in the slides and backward in the handout to page nine for those following along there.

And I will turn it back to Dr. Mikhail Menis to talk about infections following transfusion.

Mikhail Menis: Thank you, Dr. Berglund.

So transfusion-transmitted infections can be due to bacteria, virus, parasites or others transmitted through transfusion of blood and blood products – whole blood or red blood cells, plasma or platelets. It could be a bacterial infection – gram-negative or gram-positive. It could be viral transfusion-transmitted

infections such as West Nile virus, dengue, parvovirus, cytomegalovirus and others.

It could also be parasitic transfusion-transmitted infections, including malaria, Chagas or babesiosis, as well as (prime) transfusion-transmitted infections such as Creutzfeldt-Jakob disease.

So current ICD-9-CM coding of 999.3, other infection is not transfusion-specific. However, under 999.3, there is a code 999.39 that is infection following other infusion, injection, transfusion or vaccination. So we propose to make a revision to the 999.39 code and have a new code to read infection following transfusion, infusion or injection of blood and blood products.

And at a note, code first immunodeficiency virus disease so as to be according to the guidelines of ICD-9 code so that if the HIV is transfusion-transmitted, HIV would be coded first followed by 999.32 or whatever the new code is infection following transfusion, infusion or injection of blood and blood products. So that's a general scope. And we would probably propose to add a note to the human immunodeficiency virus under the code 042 as well to say you have additional code to identify infection following transfusion, infusion or injection of blood and blood products for the same reason as to record the HIV code first and then if it's transfusion-transmitted code and specific transfusion-transmitted code. So this is a general overview perhaps, Dr. Berglund.

Dr. David Berglund: OK. Dr. Menis, shall I go ahead and talk about the codes a little further?

Mikhail Menis: Sure, sure, please.

Dr. David Berglund: OK. And thank you for the presentation and the explanation about the transfusion-transmitted infections.

Again, we did have a presentation on this a year ago in September. Also, there are a lot of concerns at that time about the existing codes, notes that is, that's appeared to imply this code should potentially even perceive the code for HIV disease and there was a great deal of concern about that possibility even though it was an existing note.

So one of the proposed changes we had here was to add a code first and while it doesn't show in this slide, we would have a code first, if applicable, human immunodeficiency virus, HIV, disease for the 042. So this would then explicitly allow for the 042 code to be used first. And then we would have the 999.32 code as mentioned for infections following transfusion. And at the point 39, we would be deleting the word "transfusion" from the title there and it would just be infection following other infusion, injection or vaccination.

So we also, at 042, we propose to add a note below the HIV code. We would have that read "Use additional code, if applicable, to identify infection following transfusion, infusion or injection of blood and blood products for the 999.32 so that it would be clear that if you do know that the HIV was related to transfusion, you can use those codes together.

Now, much of the time, the codes being proposed here, the 999.32 would be intended for more immediately occurring infection. HIV, of course, is the product when – but most infections would probably be much more acute and a more immediate issue.

So this – the changes as we've done here been intended to address some of the comments and concerns that were raised when this was brought a year ago. I'd like to invite comments and see if people think that it's reasonable to make changes as we show here or if there's other remaining concerns.

Nelly?

Nelly Leon-Chisen: Question. If you intend to identify patients that develop the HIV infection after the transfusion, right? So in that case, for those patients, you would want the 042 plus the 999.32 or are you trying to say HIV patients that develop any kind of infection because, I mean, they could be already HIV-positive and they could develop infections of – related to transfusion you know infection of the central vein, catheters, whatever else they happen to have in the course of the rest of their lives.

So is this note under 042 saying because it's best to identify infection, are you talking infection in general, any type of infection or you specifically trying to link it to the HIV infection?

Mikhail Menis: Right. I think – right. So this is intended – this is Mikhail Menis. This is intended for general. And there is – currently, there is a coding – there's a note saying that use additional codes to identify specified infection such as septicemia.

I think one of the proposals that we had too is to add or other specific infection, so it is clear that it is – it could be any infection because as I presented, it could be any organism – bacteria, virus, different parasitic infections that could occur after transfusion and posed a serious hazard to a person's health. I hope I was able to answer the question.

Dr. David Berglund: This wouldn't be intended to only identify HIV. By any stretch, it's possible that someone with HIV could get a transfusion later and get some other infection. And if that were the case and they had this code at the same time, it might not be clear if that code referred to the HIV or the other infection but we don't really have any good way to deal with multiple codes and so-called grouping of codes. That's another issue.

Yes?

Jeanne Yoder: This is Jeanne Yoder from TMA. And I'm not you know right up on top of HIV, but I would think that if you had an infusion and you have the AIDS virus, you would instead use the Z08 because I wouldn't think that the HIV would be – cause some kind of a medical condition that quickly afterwards. And so I don't know why we're worried about this HIV.

I mean, 20 years ago when it was really virulent and like, bam, you got it, you got it. But now, when you've got the other strains that take longer to manifest into a full-blown HIV, I would think that if I saw this and I saw HIV first, I would think, "Oh, they had HIV and she was – of course (inaudible) got an infection and now it's even worse. But I wouldn't have expected to have a problem with this HIV as opposed to the V08.

Dr. David Berglund: Well, the intent for this would be that it could be used later also once they came down with AIDS and were using the 042 code. They could still use this code later although that would be after the transfusion by a long time so that does get awkward for tracking purposes, of course.

Yes?

Lisa Taylor: This is Lisa Taylor. So it seems to me, in that case, we would want to be using a late effect code to show we have a complication from medical care that was done in the past.

Dr. David Berglund: OK. So if it was later like a chronic problem, you'd rather see a late effect to it. Are you trying to say that instead of the way that 999.32 is currently worded for infections – any infection following a transfusion essentially, would you think we should make this acute infection and only coding acute infections that related to the transfusion? So certainly, you can get both chronic and acute types of infection.

Lisa Taylor: I felt that's what we were looking at. So if we can go back to the medical conditions that were listed …

Dr. David Berglund: Some of these that we have, OK.

Lisa Taylor: Right. So if an infusion or transfusion occurs and then the patient gets the – is it time – are we making these time-related? And if so, we need to distinguish which ones happen immediately and which ones would be a late effect from the transfusion?

Dr. David Berglund: OK. And you'd like to see this be explicitly for acute rather than chronic or long-term types of infection is what I'm getting from you?

Yes, another comment.

Jeanne Yoder: This is Jeanne Yoder again. And for instance, a prion disease, is going to take years and years to manifest.

Dr. David Berglund: Yes.

Jeanne Yoder: I mean, it takes one and then you know but – so yes. Some of those, I mean, the plague and – so if you went through, you could – well, Yersinia, isn't that the plague?

Lisa Taylor: Right.

Dr. David Berglund: Yes.

Lisa Taylor: If I want to – if, yes.

Jeanne Yoder: I mean, when you see some of these, they all manifest pretty quickly and then some of these, they're going not stand for a long time.

Mikhail Menis: If I may make a small comment, you're absolutely right. The only thing is that – I mean, usually if there is a transfusion-transmitted infection, the investigation – so if you're talking about you know a chronic type such as HIV or – it's sort of it would be investigated and if it's decided that it is transfusion-transmitted, then this is the way to identify that perhaps HIV was transfusion-related or more chronic conditions transfusion-related.

Of course, this code will capture, with a greater precision, more acute adverse events but even with those that occur you know sometime in the past, there is a way to come back and code those.

Dr. David Berglund: OK. One of the other things that I think is being raised here is the idea of potentially doing any more than one code here, one that would be acute transfusion-transmitted infection, that's potentially another that would be either a chronic or a late effect transfusion-transmitted infection.

So if you think we need potentially two separate codes to cover those, I'd certainly appreciate that. It sounds like that was at least what a couple of the comments implied and I'm getting indications of a sense that that is something people see as potentially. Do you think that would be a useful way to approach it (inaudible) at least some people showing a sense of that idea here? Anyone who'd like to comment, I invite to …

Sue Bowman: This is Sue Bowman. I guess, I'm not sure. To me, that adds more confusion to what's already being confused and then you get into the argument of you know when you use the acute one and when do you use the chronic one and …

Dr. David Berglund: OK.

Sue Bowman: … I'm not really sure it comes under what I think of as a late effect exactly because they did technically …

Dr. David Berglund: OK.

Sue Bowman: ... get the HIV-transmitted at the beginning. At that time, it's more like a disease progression or something. It's not like they suddenly developed this new condition later on. So I don't know if that would really solve it.

                I do have concerns with what it looks like from a principle. Well, the intent was that once you had any kind of HIV-positive status at all that was related to a transfusion, those codes would be linked forever. You would be assigning this 999 code for every patient encounter for the rest of their life now, and I don't know if that – how helpful or useful that really is.

Dr. David Berglund: Yes, I'm seeing other people shaking their heads that they don't like that idea anyway. So do you think we'd be better off just to have a single code for acute transfusion-transmitted infection and not one that would related at all to the chronic or longer term ones? Would that be a better approach to take? Not seeing much – well, (inaudible) on these ideas and other comments?

Female: Actually, this one is to endorse Sue's comment. I just can't see – you know if someone has HIV for the next 25 years, I can't see using this 999.32 code for the rest of this person's life for 25 years. I mean, have we looked at 909.3 late effect of complications of surgical and medical care?

                I understand your comment that it's – this is not really late effect but it is – this is defined as late effect of conditions plus – both the 996 to 999, and this would be you know within that range so maybe what we need to do is break that down to 909.3X you know for late effect or whatever term you want to

use to infuse – I know, but there's got to be a better way than using 999.32 or whatever it is for the rest of their lives.

Dr. David Berglund: OK. Yes. I think our current notes may tend to imply we should already be doing that even if we aren't.

Now, if we – I think one of the other issues here is more – I think what Dr. Menis is trying to capture with this 999.32 code is more primarily the more acute infection although I would have to defer to him on that.

Dr. Menis, could you – would you like to comment on that? How would you feel about the idea of – what if we had a separate acute transfusion-transmitted infection code, you and I had talked about that at one point during this development. Would that be something that would potentially meet the needs of the FDA in tracking the disorders that we're talking about here?

Mikhail Menis: Sure, sure. Focusing perhaps on acute and perhaps separating acute versus a more chronic condition, it's certainly definitely a possibility.

Dr. David Berglund: Yes. The chronic conditions are often tracked in other ways.

Mikhail Menis: Sure.

Dr. David Berglund: And certainly, while we haven't even mentioned, if your chronic hepatitis B and C are also huge issues for which this could potentially be a very similar concern. And some of our notes, some people may think would tend to imply that 999.39, the current code …

Mikhail Menis: Yes.

Dr. David Berglund: ... perhaps should be used first for both HIV and chronic hepatitis B and C. Whether or not that is truly intended would be another issue and we've not weighed in on that, but we leave that for another time. In any case, we'd like to try to find codes that would help for tracking these.

Again, the original intent was largely to be able to capture acute infections due to the way these were set up. The thought was it was reasonable to include the chronic ones also whether we should just forego including those and

suggest some other type of code is another issue. So we appreciate all the comments on this. Again, it is a bit complicated. We'll certainly ask for comments in writing on this.

Oh, yes. Operator, could you please check the phone lines for questions or comments from the phone lines?

Operator: If you have a question or a comment, please press star and the number one on your telephone keypad. There are no questions or comments at this time.

Dr. David Berglund: OK. Thank you, Dr. Menis, for this presentation and for participating.

Mikhail Menis: Thanks. Thank you so much.

Dr. David Berglund: All right. And Austin, I'd like to you know switch the presentation back to the other one at this point, and I'm going to move back down there. And I will turn it back to Donna or ask her where we're going next. Am I up next again? And which one am I talking about next? We'll see what we can do. I'm going to move back down this list.

OK. We will do the circulating. Next, we'll be doing circulating anticoagulant and these are in the second diagnosis agenda.

We've had a number of questions and comments over time about what we include at code 286.5, hemorrhagic disorder due to intrinsic circulating anticoagulants.

There's a number of views. In general, all of them have an abnormally-elevated activated thromboplastin time even though they had different etiologies. One of the things included here is hyperheparanemia, which is, of course, iatrogenic when people have elevated heparin level from receiving too much heparin. There are different kinds of heparin. Some of the terms used for that can vary. You have an unfractionated heparin or low molecular weight heparin, anti-IIa and anti-Xa – 10 with an X would be ways of representing this.

There's also acquired hemophilia or secondary hemophilia, where you have antibodies to one of the coagulation factors, most often coagulation factor VIII and it may also be considered autoimmune hemophilia.

Having a separate code – a specific code for acquired hemophilia would enable people to look at this and study new treatment for it's better that it would otherwise be much more difficult or potentially have been impossible.

We've also had systemic lupus erythematosus, SLE, inhibitor or lupus anticoagulant, an antibody directed against protein-phospholipids. And these antibodies usually present a risk for thromboembolic disease.

Much of the time, people may be asymptomatic with it and there can even be cases reported of bleeding. Now, that's actually much less common though and it would ordinarily imply something else was going on too.

Now, if there is a hypercoaguable state related to lupus anticoagulant, then instead of coding 286.5, code 289.81 should be used. And if you just had a lupus anticoagulant without the diagnosis, it would appropriate to use code 795.79 which is other and unspecified non-specific immunological findings.

If you do have a bleeding disorder or hemorrhage related to the lupus anticoagulant, then you would use a 286.5 code. At least, that's the intent as far as we can ascertain.

So in order to better track all these different diseases, it was requested this could be expanded and that was requested by Novo Nordisk. We have 286.5, a number of inclusion terms that we would move to the specific new codes to be created. We would create a new code 286.51, disorders due to iatrogenic anticoagulant which would include hyperheparanemia and also anti-Xa with an X and anti-IIa.

We would also create, under this proposal, a new code 286.52 for acquired hemophilia, which would include autoimmune hemophilia and autoimmune inhibitors to clotting factors, as well as secondary hemophilia. In addition, we would create a new code 286.53 for anti-phospholipid antibody with

hemorrhagic disorder. This would include lupus anticoagulant with hemorrhagic disorder and SLE inhibitor with hemorrhagic disorder.

We would explicitly exclude cases where there was anti-phospholipid antibody without a diagnosis to the 795.79 code that I mentioned earlier. We would also exclude phospholipid antibody syndrome, 289.81, or with hypercoaguable state to that code also. And we would similarly handle the lupus anticoagulant or the SLE inhibitor. Again, all of those are anti-phospholipid antibodies so this proposal would use that more general term.

And additionally, we would create a new code 286.59 for other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies or inhibitors, and other remaining inclusion terms would remain at that other code. We would also make changes at 289.81, the primary hypercoaguable state code that we have.

We currently have an inclusion for lupus anticoagulant. We would change that to be lupus anticoagulant with hypercoaguable state. And again, we would have similar exclude notes here that would make clear that if it was something like an anti-phospholipid antibody with a hemorrhagic disorder, that would go to the 286.53 or if it was a finding without diagnosis, it would go to the 795.79.

And we would have a number of other index modifications, particularly for other terms for things which are particular types of anti-phospholipid antibodies that these would be handled appropriately. And that's – I think that covers things there.

We may have – I believe there is Dr. David Cooper who is going to listen in, who had proposed this from Novo Nordisk. And, Dr. Cooper, if you are on the phone line, we'd like to invite you to comment.

Operator, could you please check the phone lines for Dr. David Cooper?

Operator: Dr. David Cooper, if you are on the line, please press star and the number one on your telephone keypad. He does not appear to be on the line.

Dr. David Berglund: All right. He may not have been able to get on at the same time. We'll certainly invite comments from Dr. Cooper in writing subsequently. And I'd like to open things up for questions here.

If we don't have questions here, operator, can you check the phone lines for any other questions or comments on this proposal?

Operator: Certainly, if you have a question or comment, please press star and the number one on your telephone keypad. There are no questions or comments at this time.

Dr. David Berglund: Thank you very much. Let me look to Donna as to which topic we wish to do next. We'll be turning it over to Beth Fisher and she will be talking about one of our topics on the diagnosis agenda part one.

Beth Fisher: Yes. If we could go to page 42 and cover the topic Reportable Malignant Skin Cancers. This came to us from the New York Cancer Registry. A request to somehow indicate those skin cancers that are reportable versus not and specifically the basal cells, squamous cells are not those that are reported. So they asked if there was a way that – to help them with their case finding of reportable cancers to be able to indicate that in a code.

And we looked at it for [ICD-]9-CM to do it by creating a code – that you know most of our codes in the cancer chapter are size-specific, not necessarily histology, so we are proposing to add to the skin cancer code, that category 173, the ability to split out the basal cell, squamous cell carcinomas of each site from other malignant neoplasms of the skin. And as indicated in the write-up that you have there, the lengthy write-up that they provided a way that they could then not have to report. A lot of this reporting now is done electronically and there's a concern that things are being reported on patients that don't have to be reported so they're trying to find a way to electronically single out the patients that do and don't have to be reported.

In [ICD-]9-CM, now there's a room to split out the codes if something is not done too much. In that chapter, we did have the Merkel cell codes a couple of years or so ago that we added and we have the neuroendocrines that we added. But for the most part in the chapter, we know that it's primarily site-specific so

we know this is a little bit of a deviation from that, but there is room to do it and we can do it a couple of ways and we can propose it to do a couple of ways. One is what you see here in the proposal is to have one code for the basal squamous cell and then one code for the other.

There was concern maybe we should have four codes at each site, one for unspecified, one for basal cell, one for squamous cell and one for other, which is also possible. We do have room in [ICD-]9-CM to do that. So I'd invite commentary now on the need for this. Maybe there are some Cancer Registry folks here in the audience and your comments about the – what we're proposing to do, how to handle it. So I invite people in the room here to comment if there's any comments. Any for or against?

I have a few ones on this output.

Operator, could I invite – if there are any comments from the phone lines?

Operator: If you have a comment, please press star and the number one on your telephone keypad? There are no comments at this time.

Beth Fisher: OK. (Inaudible) comments just curious either way about whether or not to keep the basal cells/squamous cell together versus splitting them out separately. Any value in doing one or the other?

Dr. David Berglund: I'll jump in and say I think there would be value to separating basal cell and squamous cell. I personally think that would be one of the more valuable things and those are actually, the most common cancers and they are a bit different than each other. And the way they behave clinically is different enough and these are common enough that it could be useful to separate them from the clinical standpoint.

And I think if we're going to all the work of creating these other codes, I personally think it would be better if we did separate basal cell and squamous cell and got at least some clinical value out of the changes which I – my perception was that the changes proposed here would not provide any clinical value whatsoever but if we were separating those, then it would be of some value.

Essentially, they are looking to get certain adenocarcinomas of glands separated at the fifth digit nine codes that we have proposed here. So one of the other questions I would have would be whether at the category level 173 where it mentioned that this includes malignant neoplasms of the sebaceous glands and pseudo repair it in the sweat glands, if those should actually be individually mentioned at the fifth digit nine instead and that I think that might tend to be the case.

And I'd also be somewhat concerned about whether or not the unspecified codes are being properly followed up if the cancer registry thinks we can lump it together with basal and squamous. And that, in some cases, at least unspecified may just mean they hadn't quite yet made a diagnosis which may be made as an outpatient, and I would think they should actually follow up on those cases, but I'll have to wait until we get cancer registry input on that, I imagine.

Beth Fisher: OK. Well, we invite written input as well. OK?

And Donna, I'm going to defer to you what's next. OK, certainly. Thank you.

Donna Pickett: OK. We're going to move to page 66 and the part one of the topic package, and this is an ICD-10-CM proposal. And it's a proposal being sent forward jointly by the American Academy of Pediatrics and the National Association for Children's Hospitals and Related Institutions.

Basically, what they are asking is to expand the detail at P07 and at P07 – P072 and P073 to provide more detail on gestational age completed weeks.

Dr. Linzer, I know that you're on the call-in line.

Operator, if you could have Dr. Linzer join us in the discussion. And I'm not sure if anyone from NACHRI is actually in the audience, but if you are and you would like to come forward and speak to the proposal as well, we would welcome that input as well.

Operator: Please press star-1 on your telephone keypad.

Dr. Linzer, your line is now open?

Dr. Jeffrey Linzer: Thank you very much. Did you want me to make a comment, Donna, or …

Donna Pickett: Yes. I think it would be helpful to provide a clinical background, Dr. Linzer, on the proposal. I mean, there's a lot of information here but I think, highlighting the key elements would be very useful for the audience and for those on the phone.

Dr. Jeffrey Linzer: Certainly. In the current addition of ICD-9-CM …

Donna Pickett: You're going to have to talk a little bit louder, Dr. Linzer.

Dr. Jeffrey Linzer: In the current addition of ICD-9-CM, there …

Donna Pickett: We're still having difficulty hearing you.

Dr. Jeffrey Linzer: Hang on a second.

Donna Pickett: Operator, is there some assistance that can be provided to …

Dr. Jeffrey Linzer: How about now?

Donna Pickett: Much better.

Dr. Jeffrey Linzer: OK. All right, one more time.

All right. Currently, the prematurity codes are at the two-week groupings. And in the initial proposal for 10-CM, they were actually put together in larger groupings. And after a discussion with NACHRI, we felt that it would be more important to, not only make them look more like they were a nine, but to take advantage of the space and put them by a specific gestational age based on completed weeks which is the standard way of determining maturity or prematurity of the child so that this would give us a greater granularity and more specificity as to how preterm the child is. This would be better for tracking both hospital utilization of resources and being able to track long-term outcome of complications for these children.

After review with the perinatal neonatal section, we decided that less than –
we would use 23 weeks as the breakpoint. So because of the survivability,
under 23 weeks is still quite uncommon and at 23 weeks, we're seeing more
survivability now. So that is essentially an expansion of what's currently in
nine just to allow for better tracking of these children.

Donna Pickett: Thank you, Dr. Linzer. Are there any questions from the floor on the clinical
issues and its relationship to the request for expanded detail?

John Shaw: Hi, John Shaw. And I'm wondering since the likelihood is if we're using
I[CD-]10[-CM] for a number of years and given past experience, that's likely.
This particular area does change over time.

I lost a child at 26 weeks. Twenty years later, my daughter had a child at 25
and she's fine. I'm not sure that 19 weeks or 20 weeks, at some point, may
not be the new threshold while these codes are still in use, so do we want to go
to a single digit weeks of gestation to track that over time?

Dr. Jeffrey Linzer: I think that the current science is such that 23 weeks is really most effect
break. At one time, it would have been very easy to argue 24 weeks was, but
the studies in Europe especially have shown that there is a weight and
gestational point that it's just, under the current science, is going to be too
difficult to break. Perhaps in the future, you may be able to see something
younger but the likelihood of that needing tracking under the current system is
so small that we can't propose to go in a smaller number of weeks.

Donna Pickett: Thank you, Dr. Linzer.

Do the NACHRI representative wants to have any additional comments? OK.
Thank you. I just wanted to make sure that if you had something that we
didn't overlook here.

OK. On the coding proposal, are there any specific comments on the detail
breakout at the P072 and the P073? And we have someone coming to the mic.

Jeanne Yoder: Jeanne Yoder, TMA. Something I've noticed with the mappings is there are lots and lots of codes in ten[ICD-10-CM] that don't have an NOS. And yes, you would definitely hope that this would be documented in there.

But when you're seeing something and you're trying to clump things together or whatever, sometimes an NOS is pretty important and there are just aren't very many of those in ten [ICD-10-CM].

I'm coming from, primarily, an outpatient community. I don't know that it would happen very often but it's just something to think about.

Donna Pickett: Thank you, Jeanne.

Any other comments from the audience here? OK.

Operator, are there any – oh, sorry.

Nelly Leon-Chisen: Nelly Leon-Chisen. Sorry, I'm late trying to do the math here.

Technically speaking, it shouldn't these have like 25 completed weeks but less than 24, because if you are at 27 completed weeks, you'll have completed 26, 25, 24, 23.

Donna Pickett: Dr. Linzer?

Dr. Jeffrey Linzer: How much coffee we have this morning, Nelly?

I'm not exactly sure what your question is, but when we know how many completed gestational weeks of the new born has had, I mean, that's how we age them. So if you are 30 completed weeks and three days, you are 30 completed weeks. You're not 31 completed weeks yet. Does that answer your question?

Donna Pickett: She is coming back to the microphone, Dr. Linzer.

Nelly Leon-Chisen: No, I actually understand what this is trying to do. I'm just being fictitious because we do get questions and (Anita) can tell you, we were just talking about this, we do get questions where some of our coders are a bit confused

about what does that mean, completed weeks, that you have to have completed plus one day plus you know how many days. So my recommendation was sort of putting end limits, so beginning and end limits so that it's abundantly clear which going – and I think you know if you use your common sense, you could figure it out but I can submit some proposed language to kind of figure out the beginning and end point.

And Dr. Linzer, I can certainly run that by you before I send that in.

Dr. David Berglund: Most adults give their age in completed years, although we don't commonly use the term but I think this is a common way of giving age actually. I don't know if the range is necessary. If we were going to put it, I'd be inclined to suggest it as an inclusion term rather than part of the title.

Dr. Jeffrey Linzer: Plus currently, in nine [ICD-9-CM], it will say something 24 to 25 completed weeks, so it's not necessarily a new terminology. We're just splitting it out a little for improved granularity.

Donna Pickett: We have another question or comment from the floor?

Sue Bowman: Sue Bowman. Just another thing that I will give credit to Nelly point out, but she didn't mention it because, I guess, she doesn't want to cause any more trouble.

The note that's being edited under P07.20 should probably have an NOS at the end, gestation less than 28 completed weeks, not otherwise specified because technically, all of these codes that are here are less than 28 completed weeks.

Donna Pickett: Thank you, Sue.

Dr. David Berglund: Yes.

Donna Pickett: Are there any more comments from the floor? OK.

Operator, are there any questions on the line?

Operator: If you have a question or comment, please press star and the number one on your telephone keypad. There are no questions or comments at this time.

Donna Pickett: OK. As always, we encourage you to submit your comments in writing in case you have some other thoughts on how the language might be improved for this proposal.

OK. With that, I am now going to turn the podium over to Linda Holtzman. She will do the presentation on gastroparesis, which is the ICD-10-CM proposal. And since I've messed up my pages, I don't know what page number it is but she'll know.

Dr. David Berglund: Seventy-nine.

Linda Holtzman: This is what happens when you send e-mails to NCHS. They make you stand up here and present the thing.

I was doing some coding in ICD-10[-CM] and noticed that there was not a specific code for gastroparesis in ICD-10[-CM]. So I wrote to ask about it and to suggest that one be created.

As you see in the packet, originally, ICD-9-CM did not have a code for gastroparesis and then a specific code was created in 1994. And there was a reason for that and it seemed to me that that reason was just as valid for ICD-10[-CM]

Gastroparesis is not a particularly common condition but it's a particularly miserable one. And these are patients who frequently are hospitalized. I don't know the exact statistics, because I'm not a clinician, but they do tend to have multiple hospitalizations over the course of a year and sometimes these hospitalizations can really run into lengthy stays and significant care being given.

Gastroparesis is a chronic condition of the stomach. There's abnormal motility and delayed gastric emptying. The stomach's not really able to properly macerate the food or push the bolus into the small intestine. So the patient's nutritional health really suffers.

Patients who have this condition have early satiety, bloating, epigastric and upper abdominal pain, nausea, vomiting, in fact, it's frequently characterized by just intractable nausea and vomiting. So it's really a miserable thing to live with.

These patients, because of the intractable nausea and vomiting, they tend to get a lot of dehydration and electrolyte imbalance, severe weight loss, and all kinds of nutritional compromise. That's what they wind up in a hospital a lot, unfortunately.

Now, the most common cause of gastroparesis is diabetes. And you'll see that if you have an ICD-10[-CM] book in front of you, there are codes for – there are many codes for diabetes. And then the codes of diabetes have – under the ones for diabetes with polyneuropathy, there are inclusion notes that say it include diabetic gastroparesis.

But it doesn't specify – that note alone tells you where to go but it doesn't tell you what the polyneuropathy problem is. It doesn't tell you that it's gastroparesis.

In ICD-10[-CM], there is no code – there is no specific code for gastroparesis. It's currently indexed to K31.89 which is other disorders of stomach and duodenum.

The problem for that is that if you have someone who has frequent hospitalization that do involve lengthy stay, it will be difficult for data users to identify and associate that level of utilization with just other disorder.

So the suggestion was made to, by me, to create a specific code for gastroparesis, and I don't how to – Dr. Berglund, can you just scroll this down a little bit? Thank you.

So the suggestion is to create a specific code under K31, the same category but K31.4 which would be for gastroparesis. And this would serve two purposes.

Diabetes is the most common cause. So if you have someone who has diabetic gastroparesis, you would use the diabetes code first and then there will be an additional code where you could specify what's the nature of the diabetic polyneuropathy is causing.

So you can see in the notes that an exclude two note which means that you can use the conditions together. But we have a code first note that then says use the diabetes code as the principal, and then you have a secondary code that specifies the specific condition is gastroparesis.

The K31.4 would serve another purpose. In about one-third to one-half of the cases, they never do figure out what the cause of the gastroparesis is, and in fact, we saw that yesterday when we had to add 'if known' to the note that said code first principal diagnosis such as diabetes if known or if applicable, because in up to half of the cases, they don't know.

So the K31.4 could be used as a standalone in those cases of idiopathic gastroparesis. And that's the story.

Is there anybody here from the American College of Gastroenterology who wants to comment on this? Or Nelly?

Nelly Leon-Chisen: I think Dr. Linzer was right. I mean, I don't know if it's coffee, but a couple of questions.

OK. I understand the excludes two note, but it seems a little redundant if you have a code first underlying disease note where you know it's telling you, yes, it's OK to use diabetes code. And if you do, then you use it first.

But my question is, and I didn't know if this was something that you know that exclude two note already at K31 and not under K31.4, but is there a reason why the code first noted, for example, would be only for certain types of diabetes but not like the regular type 1 and type 2 and so on because it's arranged in the parenthetical in the code first note is different.

Linda Holtzman: Yes. Actually I had suggested having them be the same. I don't know if that's a typo or – I think it's a typo. So too much coffee over there, too.

Nelly Leon-Chisen: If it is the same – OK. So if it's the same, then we don't need both note because I mean the code first note would tell you that it's OK to use the two codes and it would provide guidance in terms of the sequencing?

Linda Holtzman: It may well be that both – I think is really just a matter of wearing suspenders and a belt.

Nelly Leon-Chisen: Or is it because of the – and I'm sorry you know I didn't bring my ICD-10-CM book.

Linda Holtzman: Come on up.

Nelly Leon-Chisen: And so I'm not sure if it's because the exclude note really refers to other codes in that K31 series and the code first instruction only applies to the gastroparesis.

Linda Holtzman: Well, I'm reading the definition of an excludes two note. And it maybe that the excludes two note doesn't give sequencing.

Nelly Leon-Chisen: No. It doesn't – it doesn't.

Linda Holtzman: OK.

Nelly Leon-Chisen: And that's why I don't know what else is in that K31 range. And maybe it makes sense for those codes.

Linda Holtzman: It also includes the keep dilation of the stomach, hypertrophic pyloric stenosis, hourglass stricture of stomach, gastric diverticulum.

So I think …

Nelly Leon-Chisen: Well, then in that case in those situations if they are related to diabetes, wouldn't you still want the note to be a code first underlying diagnosis? I mean, why is diabetes with gastroparesis handle differently than diabetes with these other conditions?

Linda Holtzman: Donna says we'll take a look at that.

I guess with gastroparesis, it's because that there is such a strong association.

Nelly Leon-Chisen: And not with the other?

Linda Holtzman: And not with the other. No.

Nelly Leon-Chisen: OK. Maybe.

Linda Holtzman: And maybe the two notes are needed first to exclude two note to tell you it's OK to use them both together than the code first note to give you the sequencing instruction. But we'll take a look at that.

Sue Bowman: Yes, and actually why it didn't support, Nelly said. Because when I looked at it, I thought having the two notes made it more confusing, not clarifying, because I was trying to figure out now what does the exclude note trying to tell me, is it the difference than what the code first note is trying to tell me.

Linda Holtzman: OK.

Sue Bowman: And it causes more confusion in my mind.

Linda Holtzman: It is different. I think part of it may be due to just you know all of us becoming familiar with ICD-10[-CM], but it really is different. The excludes note – the exclude two note tells you that you can use both codes together, then the code first note tells you the sequencing. So …

Sue Bowman: It's on your first (inaudible).

Donna Pickett: Right. Yes. We'll take a look at the note. The excludes two note was already there, so that was the only reason that I brought it forward so that we could generate this kind of discussion, because that note was already there. It's not something we added for this proposal.

And as Linda has just indicated, when you send proposals, be prepared to actually have to present them.

OK. I'd like to turn the podium over now to Beth Fisher for the other gestational age proposal, but this one is related to – in pregnancy.

Beth Fisher: OK. We're on page 63 and I guess is should check if Sean Currigan maybe on the line, (Donna Taylor) – operator, could you see if Sean Currigan is on the phone and maybe he can be patched through to us.

Operator: Mr. Currigan, if you are on the line, please press star and the number one on your telephone keypad.

Beth Fisher: Well, he's in travel. So I mean he may not be available.

Operator: He does not appear to be on the line.

Beth Fisher: Or I guess if he uses our speaker dial-in line, you just have to unmute his line.

Anyway, I'll go ahead and do this presentation. I'm sure everybody's fine. We'd like to have him.

This came to us through American College of OB-GYN through work with physicians' consortium of a variety of group put together you can see in your packet there in the first paragraph kind of describes that.

And you know we started working. And as Donna said yesterday when somebody said why don't we wait until ICD-11. She went through the history of where ICD-10 became ICD-10-CM and how long it's been and it's – we know it's been a while.

So back in 1994 when we were first working on [ICD-]10-CM before my life in the government, so – it was requested by ACOG at that time that said it would useful to have the ability to track things by trimester instead of the fifth digits where right now in 9 and 10, we track by whether you deliver antipartum, postpartum.

So we did that with the whole chapter. And then they came to us this summer and said, "We'd really like to know the weeks and the day, too."

So it's quite – it'll be quite difficult to go back to chapter 15 and do that to all those codes really. It would just be a major undertaking. It might delay the whole process again.

But they have interest in tracking more detailed information about the gestation of the pregnancy when something – complication, if something occurs to be able to know the specific weeks.

And so we – initially, they want us to know the weeks and the days and also to be able to track in the code how that was determined or if by ultrasound or by menstrual period. And that's really got to be difficult to even think through.

So kind of scaled it back and said, "You know, maybe if we first start with tracking the specific weeks and do it through a Z-code," which is the Z-code. That's the last chapter in 10-CM.

So what we were proposing – and we try to leave room for the future if we were to expand this further.

In the discussions with the ACOG physicians, even there, we sort of – when we got down to the days, I guess it was sort of a discussion about at this point in time what would you do with that information.

So we scaled this back to do it by weeks only through a Z-code. And so what we're proposing is that in chapter 15, we'd still have our codes broken out by trimester. As you see, we've got the notes there printed on page 63 that appear right now in ICD-10-CM. And then we proposed creating a new category in the Z chapter, Z35, which would provide the additional information.

So if you have a first trimester patient then you would come to Z35 and indicate the weeks of gestation for that.

And then we broken it out – we broke it out further into specifics. We kind of grouped in the first trimester of zero through seven, eight through fourteen right now indicating – maybe that's probably – well, that it may not be necessary to have the individual weeks, but there is room to do that.

And then in the trimesters, we broke it out week by week all the way up through the 43rd week thinking that it should be enough.

If we broke this out by days of the week, then you end up with about 300 codes, so – 270 or 300 codes. So that's why we sort of said, "Wait a minute. Let's just start with the weeks."

But – so the idea is that you would code first week or I mean you would use a code in chapter 15, and that indicates through the fifth or sixth digit depending on the code you're using which trimester you're in. And then, you would come to this category to indicate specifically the week.

And I have a feeling over Nelly's question earlier maybe they're going to wonder about completed weeks. But my understanding is the week, and I could get qualification on this but the week is – if you're any point in time during that week, between day zero and day six or day seven I guess.

Nelly Leon-Chisen: Nelly Leon-Chisen. I just want to clarify. I don't have the same question …

Beth Fisher: OK.

Nelly Leon-Chisen: ... about the number that we get, because I think on page 63, they did a nice job.

Beth Fisher: OK.

Nelly Leon-Chisen: That's the kind of thing I was talking about where it says less 10 weeks you know 14 weeks zero days – 14 weeks zero days to less than zero days.

Beth Fisher: There should be.

Nelly Leon-Chisen: I'll give you a beginning and an end point.

Beth Fisher: All right. I didn't mean to single you out.

Nelly Leon-Chisen: Well, whatever. I mean that kind of – I commend that, and you're right.

Beth Fisher: OK.

Nelly Leon-Chisen: Everybody knows it needs to be transferred over to the Z-codes as well. Yes, it's in a different section.

Beth Fisher: OK.

Nelly Leon-Chisen: Yes.

Beth Fisher: OK. So I would invite in the room any comments on this proposal. OK.

Kathy Myrick: Kathy Myrick. Would we put this additional code on every patient visit into the clinic not just hospital?

Beth Fisher: Well I suppose there's a potential for that. Yes.

                    OK. Any other comments in the room? If not I'd like to invite if there's any comments on the telephone, operator?

Operator: If you have a comment or a question, please press star and the number one on your telephone keypad.

                    And we have Dr. Linzer on the line.

Beth Fisher: Hello, Dr. Linzer.

Dr. Jeffrey Linzer: Hello. I have a question, and more for conservation of space than anything else and I'm not sure if the 10 convention would allow it.

                    But is it possible to just use, for example, the Z35.1 and take it out to three digits from the decimal so that you could fit all of the trimester information under one subcategory instead of using three subcategories or four subcategories.

Beth Fisher: Well, I guess what we're trying to – we were trying to leave room for the potential of days in the future. I'm not – so you – or is this ICD-10-CM you're trying to save space?

No. OK. So you were just wondering if you could fit it all like Z35.1 would be for the first trimester, 0.2 for the second, and 0.3 for the third. Is that what you're …

Dr. Jeffrey Linzer: No. I was just wondering why you couldn't leave it all at Z35.1, take it out far enough bit. You could just use one set, subcategory, to catch it all.

Certainly, what the alternative would be just to use 0.1, 0.2, and 0.3 so that you could separate up the trimesters in that way. But I think to take so many subcategories just knowing what's happened in ninth, maybe an unnecessary waste of space.

Beth Fisher: OK. Well, we – I mean, OK. We can – we think it, but yes, space concerns.

OK. Are there comments here in the room?

Pamela Ruebelmann: My name is Pamela Ruebelmann from Clarity Coding. I just – I'm very novice to ICD-10, so just bear with me. But I'm reading a note [that] says third trimester is at 28 weeks, zero days. So I'm reading that as 28 weeks. And when you look at 35.37 because it goes to second trimester and not the third trimester. So am I interpreting this the 28 weeks is actually a second trimester or a third trimester?

Beth Fisher: OK. I see what you're saying.

Pamela Ruebelmann: Yes.

Beth Fisher: OK. I have to read it myself. Twenty-eight weeks, zero days. OK. Looks like I need to look at that.

OK. Any other comments in the room?

OK. Next topic.

Pardon?

Were there any other comments on the phone, operator?

Operator: There are no further comments at this time.

Beth Fisher: OK. Then we'll move with David here in control.

Dr. David Berglund: All right. We've got a number of interstitial lung diseases.

Is Dr. Lawrence Nogge from John Hopkins here? Yes. We're expecting Dr. Francis McCormack I believe – oh, you're here, too. All right.

Dr. McCormack, would you like to talk about any of these. Shall I start by doing presentation on the interstitial lung diseases of childhood and then, Dr. McCormack, do you have anything you'd like to present?

Dr. McCormack, did you have any PowerPoint slides? You do? OK. Do you have those on something just so that we could give to (Austin), the gentleman who is in the room right here? Let's try and do that.

And I am going to – let's see. I will – Dr. Nogge, did you want to present or do you want me to present on the childhood interstitial lung diseases? I think you wanted me to present on that and then you could comment after. That'll be fine.

And we had originally been thinking we might do more of these this afternoon and we may have to break partway through Dr. McCormack's presentations later, but we will probably handle them in that way if we can.

I'll go ahead and talk now about the interstitial lung diseases of childhood and we'll move on from there.

We'll start with neuroendocrine cell hyperplasia of infancy. This is one of interstitial lung diseases that occurs in children. We'll be hearing more about a number of other interstitial lung diseases, but these are unique to children. And this is one disease that can cause need for oxygen use that they may need for a number of years, and it's being diagnosed more widely.

Lung biopsy is needed to make the diagnosis based on histological staining of the neuroendocrine cell.

While as I understand the underlying cause isn't elucidated as yet, there is some familial incidence, meaning that it could very well be genetic, although that's not yet understood. At this time, we don't have a specific code for this or even an index entry.

The American Thoracic Society and American College of Chest Physicians have requested creation of specific code for neuroendocrine cell hyperplasia of infancy along with a number of the other interstitial lung diseases that we'll be covering a little later here. And we will cover those in more detail. I'm just going to go through each of these and then invite comment.

Another of this interstitial lung disease is pulmonary interstitial glycogenosis. And this is a disorder that's found often in newborns that have hypoxemia that maybe out of proportion to the other finding.

There's thickening of the alveolar interstitium, proliferation of a poorly defined clear cell population seem to be causing this and it causes some marked diffusion abnormalities for oxygen. So basically these infants cannot oxygenate their blood as well as they should be able to.

You know, it can occur as the primary finding or it can occur along with premature lung disease or with congenital heart disease. And it is a major problem for those infants that have it. Again, in disorder, a lung biopsy is required to establish the diagnosis.

You know, again, in ICD-9-CM, we don't have even an index entry for this. I will also comment it's unrelated to the glycogen storage disease coded at 271.0 glycogenosis.

Coders might tend to think it should go to that code since there is the word "glycogenosis" mentioned with it. But it is really completely unrelated and it wouldn't be appropriate to use that index entry. So ATS and ACCP again requested a specific code for pulmonary interstitial glycogenosis.

Moving on, the surfactant mutations of the lung are actually a group of disorders that do cause significant morbidity and mortality for children. These

are one of the bigger reasons for pediatric lung transplants. They may present in a newborn period or later childhood with an unknown product lung disease.

There are a number of different surfactant mutations each that are characteristic and individual and have their own course and their own prognosis. And ATS ACCP did suggest that it's maybe most beneficial for each of these to have their own code or failing that to at least create a single code for surfactant mutations of the lung.

Now, there's also some growing evidence that such mutations may impact on other lung diseases. They may be gene modifiers for other types of lung disease. Surfactant is very important for lung homeostasis for normal functioning of the lung and so a mutation in one of these genes may impact on other disease then.

So having specific codes for these or at least for surfactant mutations generally would give a help in tracking these and identifying them and enable things to be handled with them.

Specific surfactant mutations include surfactant protein B mutations of the lung, surfactant protein-C mutations of the lung, surfactant associated ATP-binding cassette A3 mutations of the lung, and surfactant associated thyroid transcription factor 1 mutations of the lung. And they do have particular abbreviations for each. ATS and ACCP request creation for specific codes for this again.

Another disorder in interstitial lung is the alveolar capillary dysplasia with vein misalignment, abbreviated ACDMPV. It's a developmental lung disorder. Children present immediately postnatal with a rapidly progressive respiratory failure.

They have severe pulmonary hypertension that progresses to death within two months of life and that's despite therapeutic intervention for the pulmonary hypertension ventilation strategies and extracorporeal membrane oxygenation or ECMO. In general, all of these fail to help these infants.

They're having cases of family history and that would suggest there may be some type of an autosomal recessive gene involved although the cause of this is still under study.

It is first under recognized, it is diagnosed by lung biopsy or on postmortem evaluation. And again, we had a request for specific diagnosis for that.

We have two options noted here. Option one would create a set of codes with a new subcategory of 516.6 for interstitial lung diseases of childhood. We will be covering the other – some other proposed codes using other interim numbers for those wondering what happened to the other codes of 516 that we come before this, why we're using 0.6. We'll get to those later.

The 516.61 would be neuroendocrine cell hyperplasia of infancy, 516.62 pulmonary interstitial glycogenosis, 516.63 surfactant mutations of the lung, and 516.64 alveolar capillary dysplasia with vein misalignment. We would also create a new code of 516.69 for other interstitial diseases of childhood under this proposal.

We also have a second proposal option two with the idea being to break out the surfactant mutations of the lung. And in this case, we would again have 0.61 for the neuroendocrine cell hyperplasia of infancy, 0.62 pulmonary interstitial glycogenosis, and at this time we would have 516.63 surfactant protein B mutations of the lung, 0.64 surfactant protein-C mutations of the lung, 0.65 surfactant associated ATP-binding cassette A3 mutations of the lung, 0.66 surfactant associated thyroid transcription factor 1 mutations of the lung, 0.67 would be alveolar capillary dysplasia with vein misalignment, and 0.69 would be other interstitial lung diseases of childhood.

I'd like to invite Dr. Nogge to comment. Yes. Let's let you comment on these.

Dr. Lawrence Nogge: Sure. Lawrence Nogge from John Hopkins representing ATS. I'd really like to advocate that.

People may be wondering why there need to be separate codes for children in the scheme that Dr. McCormack is going to present, those diagnoses really do not apply to children.

For example, the most common diagnosis, UIP, usual interstitial pneumonia doesn't occur at children in less than 10 and some of the other histologic diagnoses have very different etiologies and courses.

Secondly, there are no good codes that describe these. Also, my colleagues, they're using very non-specific codes such as hypoxemia of the newborn, the code for bronchopulmonary dysplasia 770.7, I believe, which is really designed for lung disease following premature birth.

There's 515, which is for pulmonary fibrosis following inflammation, although inflammation isn't part of the course of many of these and in fact, many of the kids, particularly with knee high don't have fibrosis.

So it's been a real problem for the physicians and hospitals caring for these children to try and decide how to code them and be able to track them appropriately.

Dr. David Berglund: OK. I appreciate it. Oh, Dr. Nogge, when I look at these, it looks like at least some of the other disorders that Dr. McCormack's going to talk about later probably do occur in children although the most, at least certain ones don't, and that may not be as clear on which one do occur in childhood and which one don't.

I would imagine for those that do, we could probably use other proposed codes, too. But there's at least some that this would definitely apply to.

Dr. Lawrence Nogge: But some of the ones from the literature seem to occur in children, for example, desquamative interstitial pneumonitis. When that is seen in children, that is due surfactant mutations of the lung. We know that now.

So that's really a separate entity and that's part of the reason for proposing the separate code to be more precise…

Dr. David Berglund: OK. So we might want to exclude, if it was from the proposed (clinician) codes that we might be proposing, it might be appropriate to exclude surfactant mutation in children to these, I take it?

Dr. Lawrence Nogge: Correct.

Dr. David Berglund: So that would – OK. That's not something we have done at this point as yet, and we hadn't really discussed this in as much detail, but I definitely appreciate that aspect of things. So that's something we may want to add then.

Dr. Lawrence Nogge: OK, great. Thank you.

Dr. David Berglund: Thank you.

Do we have other comments here? Yes?

Jeanne Yoder: I was really hard for an NOS on this one. Oh, shoot.

Dr. David Berglund: Sorry. Jeanne Yoder mentioned she looked really hard for an NOS on this and couldn't find one. Is that the idea? OK. I summarized your comment. Thank you, Jeanne Yoder.

Kathy Rivera: Kathy Rivera. I do a lot of work for Children's Hospital. And the coders ask the children's facilities, really are looking for the physician to say childhood or they will not assign these codes.

We find this with asthma, currently because this childhood asthma, I don't care if this child is three, the coders don't have those words, and they're not assigning the childhood code. Can't convince them to do otherwise because they feel they don't have the documentation of the actual childhood.

So with these, without having an age to base it on, expecting physician documentation that isn't going to happen because it should be natural to say 6-year-old, it's a child. But there are a lot of coders out there that will not use those codes because they don't have anything more than not otherwise specified. I don't know if we can, in some way, define childhood a little more clearer.

So you know each facility can write their own policies and procedures relating to that, but then you can define those very differently across the nation, which wouldn't help in any kind of consistency. And I just throw asthma out there because it's really one of the ones that if they're 15, is it a child or – it's very difficult.

Right. Right. I'm with you. But I just want to throw that as a concern because I can see that saying you know that the coders don't want to say any interpretative just because they know the patient's age and they know the diagnosis that they can relate those.

Dr. David Berglund: OK. So that is certainly something for us to consider. I'm not sure how we might modify things in dealing with that, although there are certainly certain diseases that do occur in younger children that don't occur in adults. And that's what we're trying to convey here.

Another comment.

Linda Holtzman: I guess this follows on with Kathy's comment.

Dr. David Berglund: Linda Holtzman.

Linda Holtzman: Thank you for identifying me.

Just a follow on to Kathy's comment, I confess I've never heard any of these conditions and I've never coded them. But from what written here, most of them seem to be specifically of newborn or they're identified in the course of the newborn treatment or the immediate neonatal period, most commonly present in the newborn period. So I guess what I'm wondering is why aren't these in chapter 15?

I know you must have thought about that so I'm just curious what the reasoning was.

Dr. David Berglund: Well, that's certainly an excellent question. We could contemplate that. But they can occur somewhat later, too.

Dr. Nogge, would like to comment on that?

Dr. Lawrence Nogge: Yes. That maybe appropriate for the alveolar capillary dysplasia which is almost exclusively to this and newborns.

These codes are really being driven by pediatric pulmonologists that were seen in the children at a later time. So – and in fact, while we're trying to distinguish from adult, there are adults who can have these disorders as well. But I think this is the best place for them in terms of – that these physicians being able to find them and act appropriately.

Dr. David Berglund: But some of these at least can occur in a little later childhood not just in neonates that which – that's part of whether here mainly?

Linda Holtzman: As a clinical question, do these originate in this perinatal period and then they're identified later in life or do they originate later in life that actually gets to a coding point?

Dr. Lawrence Nogge: It depends on which of the diseases. Some of them start right at birth. Others really don't manifest until later in life.

Linda Holtzman: Are they associated with conditions, adverse, or during the neonatal period and then they're identified later on?

The reason I ask is because if the physician may – you commented the physician would look to see it in this part of the book, but as a coder, if I'm looking at somebody who's 14 days old or five days old or even somebody who's 15 years old but they tell me that because of something that happened during the birth process, then I'm going to be looking in chapter 15, the neonatal codes.

So you know I certainly think we need for codes for this. It's just a question of where to put them so people can find them most easily.

Dr. Lawrence Nogge: I mean one could obviously argue that in, for example, the surfactant mutations that since they're a genetic disease, they're present at birth. The physical manifestations and symptoms may not present for quite some time

after birth. So there's some variability and I think we don't know mechanistically for the other ones. So it's somewhat disease entity specific.

Dr. David Berglund: I guess that would raise the question of whether we should put them in the congenital section as oppose to the perinatal section, but that's another issue entirely of that. Thank you.

Another comment.

Kim Dura: This is Kim Dura. I have a follow-up, I guess, a clinical question. Do these conditions resolve during childhood or do they continue beyond childhood?

Dr. Lawrence Nogge: That depends on the condition. I have a slide which I can show you. And some of these are ultimately fatal in the childhood period, particularly, for example, alveolar capillary dysplasia. Others persist throughout life. We think that neuroendocrine cell hyperplasia, the course is gradual improvement, but time period for resolution is years.

Dr. David Berglund: OK. Operator, can you check the phone lines for comments or questions. I think Dr. Linzer maybe wanting to comment.

Operator: Yes, Dr. Linzer, your line is now open.

Dr. Jeffrey Linzer: Thank you very much. First of all, let me congratulate the coder for not using the childhood asthma code. If we could ever get done where it showed, replace the extrinsic and intrinsic term I think it would make everybody happy.

I do have a question for Dr. Nogge if he had discussed this proposal or reviewed it at all with the perinatal section of the academy.

Dr. Lawrence Nogge: No. To my knowledge, that hasn't happened.

Dr. Jeffrey Linzer: Yes. I have not had anything from either the section of pulmonology or the perinatal section from the academy come through on these. They certainly are interesting.

I've spoken with some of the pulmonology colleagues here in Atlanta about some of these conditions. But I think some of the issues about where they may sit in the book that have raised need to be addressed.

And certainly, I'm going to have to review this with my perinatal colleagues and the pulmonology section before we can make any comments one way or the other about them.

Dr. David Berglund: OK. Well, yes, thank you, Dr. Linzer. Any other comments on that? I know these are fairly involved.

Lisa Taylor: This is Lisa Taylor. I just want to second that. I have a concern about where these belong in the book.

As a professor of coding, I do teach my students that those codes in chapter 15 can be used later in life if they originated during the perinatal period. So I think that's really important here.

Dr. David Berglund: OK. Thank you. We will look further at that, certainly.

Do we have any – operator, could you please check whether there are any other questions on the phone lines?

Operator: There are no other questions at this time.

Dr. David Berglund: OK. Thank you to Dr. Nogge and thank you to Dr. Linzer for commenting and for all the comments on this.

I would like to move next to some of the other interstitial lung diseases that Dr. McCormack is going to present on.

Dr. McCormack, did Austin have your slides? Let's have you come up and we can have you present up here. Good.

Thank you, Austin. And Dr. McCormack, I'll turn things over to you.

Dr. Francis McCormack: So I'm here in behalf of the American Thoracic Society and the American College of Chest Physicians to advocate for new code for the

interstitial, several different interstitial pneumonias and for two special diseases in a separate category called adult langerhans-cell histiocytosis and also lymphangioma myomatosis.

And Dr. Berglund, do you have a preference for which we talk about first?

Dr. David Berglund: Go ahead and take them in order. You got them in your slides. That's fine.

Dr. Francis McCormack: In terms of the idiopathic interstitial pneumonia – I don't know if have the – can we go forward? This is a set of lung diseases. I don't have a pointer here but they are characterized by scarring.

Dr. David Berglund: I think there's a pointer right here.

Dr. Francis McCormack: Thank you.

These are set of diffuse parenchymal lung diseases that can be subdivided into category that include dose to the drug and cause vascular diseases, the idiopathic interstitial pneumonia, which I'll talk about first. The granulomatous diffuse pulmonary parenchymal lung diseases such as sarcoidosis and other forms of diffuse parenchymal lung diseases including lymphangioma myomatosis and pulmonary langerhans cell histiocytosis.

We'll talk first about the idiopathic interstitial pneumonia which are really divided into idiopathic pulmonary fibrosis and other idiopathic interstitial pneumonias including desquamative interstitial pneumonia, respiratory bronchiolitis interstitial lung disease, acute interstitial pneumonia, cryptogenic organizing pneumonia, nonspecific interstitial pneumonia, and lymphocytic interstitial pneumonia.

These diseases present in similar ways, but they have very distinct pathologies. That's redundant, I think. These diseases can present in both an idiopathic form and also the same pathology – lung pathologies can present as a component of collagen vascular diseases including rheumatoid arthritis, systemic lupus, mixed connective tissue disease, systemic sclerosis,

polymyositis, dermatomyositis, Sjogren's disease, and undifferentiated connective tissue disease.

These pathologies that I mentioned earlier, usual interstitial pneumonia, nonspecific interstitial pneumonia, lymphocytic interstitial pneumonia, cryptogenic organizing pneumonia, and obliterative bronchiolitis can be seen in each of these collagen vascular diseases to different extents.

Usual interstitial pneumonia is the most common pathology seen in rheumatoid arthritis and nonspecific interstitial pneumonia is a secondary pathology. That order is reversed for other interstitial diseases such as mixed connective tissue disease where nonspecific interstitial pneumonia is the most common pathology and usual interstitial pneumonia is the less common one.

The other important interstitial lung diseases that I'll mention are pulmonary Langerhans cell histiocytosis and Lymphangioma myomatosis. These others already have code.

This is the proposed codes and Dr. Berglund could probably do a better job at this that I will, but – and all of these diseases were formally coded under 516, other alveolar and parieto-alveolar pneumopathy, but it didn't distinguish them and it wasn't optimal for clinical research in tracking the diseases, the clinical effectiveness research et cetera.

The 516.0 is pulmonary alveolar proteinosis, it was a preexisting code; 516.1 idiopathic pulmonary hemosiderosis; 516.2 pulmonary alveolar microlithiasis. And we're proposing to extend the 516.3 category which I believe was idiopathic fibrosing alveolitis – is that true – to include all of the idiopathic interstitial pneumonias including idiopathic interstitial pneumonia category non-otherwise specified.

Idiopathic pulmonary fibrosis under 516.31, idiopathic nonspecific interstitial pneumonitis, acute interstitial pneumonia, respiratory bronchiolitis interstitial lung disease, idiopathic lymphoid interstitial pneumonia, desquamative interstitial pneumonia, and cryptogenic organizing pneumonia and then in category 516.4 Lymphangioma myomatosis and 516.5 Pulmonary Langerhans cell histiocytosis.

And Dr. Berglund and I had some late e-mail exchanges last night, and I don't think – I think he fell asleep before I did. So we didn't quite get to this.

But I'd also like to propose that we consider categorizing the lung pathology who can present with these connective tissue diseases under 517, lung involvement and diseases classified elsewhere. This lets each of the lung pathology that are commonly seen in the connective tissue diseases, and I imagine that the optimal coding then would be for the underlying collagen vascular disease or connective tissue disease to be coded first and then each of these pathology or the pathologies that are relevant to the interstitial pneumonia in that disease could be coded as a secondary code.

Again, this is all late night musing but I think it would be very helpful to us as clinicians to be able to indicate exactly what pathology we're seeing in each of this connective tissue diseases.

Dr. David Berglund: This one was not in our handouts. We will have to look at it a little further, I think. This one basically would get to the pathological or histopathology that we would be on things like volume biopsy.

We'll have to probably look at this a little more. We do have existing other codes at 517 that would be different from this. So we would have to deal with it in a little different way than what we have seen here. But we will – we'll certainly take this into consideration in sorting out how to handle things.

Dr. Francis McCormack: Just to address the issue of the existing codes under 517, one was rheumatic pneumonia, and in fact, that's the one manifestation of lung scarring and rheumatoid arthritis which can be due to anyone of the number of five different pathologies. So I think that's an outdated term.

And then second one was lung diseases associated with systemic sclerosis that I remember and, again, there are many pathologies that can be associated with systemic sclerosis. So that code is a little broad. But I do acknowledge that you know we need to think carefully about how this would best be put together.

So I'd be happy to go into any more detail in any of these interstitial lung diseases if that's helpful.

Dr. David Berglund: Let's go ahead and take questions on.

Dr. David Berglund: We got a question right here.

Female: I have a clinical question regarding this code sec. You said we're under 517 now and had certain histopathologies. Are those histopathologies the ones that are listed here?

Dr. David Berglund: Yes, the histopathologies are the existing codes are a bit different. And he just mentioned them verbally at some of the things we have there. But the histopathologies that we had here – the histopathologies on the other slides were basically ones that can be found in many of the other disorders that we had at 516

Dr. Francis McCormack: Right. So – I left out the third number there but in any given disease such as rheumatoid arthritis, any one of a number of pathologies can be seen.

Dr. David Berglund: Yes, 517.1, we have an existing code rheumatic pneumonia which would, of course, be with the rheumatoid arthritis.

So we wouldn't be able to use the code 517.1 for the other histopathology that you showed in the other slides, but we can contemplate potential ways of including that information in some other way.

We'll have to look at that further.

Dr. Francis McCormack: OK. Did you want to discuss lymphangioma myomatosis at all, Dr. Berglund, because of the neoplasm versus interstitial lung disease aspect of that?

Dr. David Berglund: Certainly. Shall I pull up what we have on that? Let's – Austin could you switch to the other display? Here we go.

Lymphangioma myomatosis or LAM. We had talked about this a few times earlier, Dr. McCormack and I talked that is. And it is in some ways like a neoplasm. And one of the problems at this point is now it is rare, it is frequently fatal, it almost only just affects women, it does involve infiltration of the lung with neoplastic smooth muscle cells and then cystic destruction of the lung tissue.

Right now, coding in ICD-9-CM, it's sometimes maybe coded to 171.9, from what Dr. McCormack said. That's commonly used by clinicians. From our indexing, I think it would lead people to use code 238.1, which is neoplasm of uncertain behavior of other nonspecified sites and tissue. Having a more specific code would have a lot of benefits to it in being able to track it better.

And clinically from what I understand from you, Dr. McCormack, it's much more consistent in its behavior with an interstitial lung disease not so much in neoplasm even though the tissue cells maybe neoplastic in nature also.

So we have the request that it be instead coded to category 516 at a proposed new code – 516.4 as was shown.

Would you like to say anything further about it?

Dr. Francis McCormack: That's really the issue.

Dr. David Berglund: Summarizing it briefly?

Any comments on that one? I recognize this one would be moving something around which might create other concerns. So I certainly would like to invite comments on LAM specifically. Would anyone like to come to the podium and – or come up to the mic here and comment on it?

It's OK. I know we're going to break for lunch soon but we still got a few minutes left.

If we don't have anyone coming up, operator, would you like to check the phone lines for any comments at this time?

Operator: Certainly. If you'd like to ask a question or if you have a comment, please press star then the number one on your telephone keypad.

We have the line of Linda Small. Your line is now open.

Linda Small, your line is now open.

I'm sorry. She's no longer on the line.

Dr. David Berglund: All right. We'll come back to the phone line in a moment. And we now have a comment here from Nelly Leon-Chisen.

Nelly Leon-Chisen: Yes, hi. I understand your interest in having a specific code to identify this condition, but it's still related to neoplasm, right? It's a neoplastic tissue growing. Why did you want it in this section as oppose to just creating a unique specific code in the neoplasm section?

Dr. Francis McCormack: Right. So it is a disease that's taken care of physicians who are expert in interstitial lung disease. If you were to poll the community of pulmonologist and ask a question about whether this is a neoplasm, I think the answer would be no toward the 80 percent end of the spectrum. It's only through recent revelation at the molecular level that it appears that this is extremely low grade neoplasm at the extremely mild end of the spectrum.

So these are smooth muscle cells that appear to migrate to the lung. They appear to be clonal there. So it is just becoming more apparent, but it's not yet widely adopted by the pulmonary community that this entity has interstitial lung disease characteristics but a neoplastic underpinnings.

So I think that it's more appropriately coded as an interstitial disease. I think the evolving science suggests we're going to come to believe it's a low grade neoplasm. But I don't think it's widely accepted yet.

Dr. David Berglund: And as I understand, it's not treated by oncologist using traditional methods of treating neoplasms either and it involves cystic destruction of lung tissue rather than growth in the lung as most types of lung neoplasms cause and again, it's neoplastic smooth muscle cells.

Now, how and why they come there whether it was it going to be a secondary neoplasm isn't clear. While it does seem to be somewhat neoplastic, it's not really clear. It's not a primary lung cancer, but it's not like most secondary lung metastases would be.

So it's really quite different clinically from most cancers that we would find in the lung, either primary or secondary cancers. So clinically, it doesn't behave like what we think of as a cancer of the lung or – either metastatic or primary.

So for that reason, it seems it would really just create confusion and potentially maybe creating confusion to be coding it that way.

And there's – for that matter, there's probably not other room to expand it within the neoplasm chapter that and particularly the behavior being quite different from most neoplasms, it seems problematic to try to continue coding it there in some respects.

Linda Holtzman?

Linda Holtzman: Dr. Berglund, can you just go back to the previous slide?

Dr. David Berglund: That one?

Linda Holtzman: No, the previous one.

Dr. David Berglund: Oh, the slides of Dr. McCormack again.

Dr. David Berglund: You mean the slide that was on the white background or the blue background?

Linda Holtzman: No, the blue background.

Dr. David Berglund: Blue background. Austin, could you bring up the other slides again?

Which one do you want? You want the one that showed 516? You probably can back it up using Back. Here we go? That one?

Linda Holtzman: Yes. I might have missed it in the packet but I don't see 516.4 or 516.5, and 516.7 in the packet. I see everything broken out under 516.30.

Dr. David Berglund: Oh, those? The other ones are actually in the other part of the packet unfortunately.

Linda Holtzman: OK.

Dr. David Berglund: The lymphangioleiomyomatosis – let me say that a little more slowly – is on page 23 of your packet. So that's one that we've talked about. And the adult pulmonary Langerhans cell histiocytosis is on page 28.

I'm getting my tongue tangled. There's something here, I'm afraid.

OK. So my apologies. We do have things a bit scattered around there. And we will want to invite more questions on these. – yes?

Kathy Rivera: I have one because with the potential of seeing a neoplasm associated, will this then include the neoplasm that you would omit a neoplasm code if that was identified – the LAM? It's kind of – because they're uniquely different?

Dr. David Berglund: Yes. I think it would include the neoplasm again. While we have told people to use the neoplasm code based on the indexing, it really doesn't act like a neoplasm.

So yes, this would include the neoplasm, it would include basically the entire disease as a new code. Yes.

All right. Let's – Austin, could you switch back to the other – what we were jumping around a bit I recognize here.

And I will comment, we are going to lose the recording and phone precisely at 12:30. So we will need to break for lunch then.

After lunch, we had a number of speakers who wanted to participate. And we had some who were going to be participating from remote locations that want to be on right then.

So I apologize, Dr. McCormack, but we may actually not come back to this immediately after lunch. We may end up having break and then come back to it after that.

This has gotten a little awkward in sorting out logistics but we will do our best to do what we can with it.

In the meantime, let's skip to what we've got on screen now – adult pulmonary Langerhans cell histiocytosis. And again, this is another rare interstitial lung disorder, unknown etiology. It occurs almost exclusively in smokers. Peak incidence age 20 to 40 years, completed years. And it is generally a single system disease with the so-called Langerhans cell granulomas that infiltrate and destroy the distal bronchiole.

There's a typical high risk PT pattern with some small nodules with or without cavitation, some cyst in the upper lungs generally. It's somewhat distinct from multisystem histiocytic disorders that mainly occur in children and adolescent where the pulmonary disease is rarely do focus.

Now I think in histiocytosis, if the pulmonary disease can occur some – is that correct, Dr. McCormack?

Dr. Francis McCormack: In adult pulmonary Langerhans cell histiocytosis you can have bone involvement and skin involvement, but very uncommon.

Dr. David Berglund: OK. So you can actually have the bone and skin involvement in this one also.

Now, we don't currently index adult pulmonary Langerhans cell histiocytosis in [ICD-]9-CM. We do have histiocytosis. We've got its code 277.89, other specified disorders of metabolism. We have some of those. And also histiocytosis X and acute differentiated progressive histiocytosis are coded to 202.5 which is Letterer-Siwe disease, and I guess that's widely considered the more pediatric form of histiocytosis.

It's potentially the case people would use one of those two codes 277.89 or 202.5, but it's a bit different with the interstitial lung involvement American

Thoracic Society and ACCP would currently want to be using code 516.8 or that, and they're proposing giving it its own code. Its clinical behavior is consistent with an interstitial lung disease more so than either a neoplasm or metabolic disorder.

So we'd be proposing to exclude this from the other codes and instead coded to a new code – 516.5 for adult pulmonary Langerhans cell histiocytosis.

And Dr. McCormack, any other comments on this?

Dr. Francis McCormack: No.

Dr. David Berglund: OK. Any comments from the audience on this? Anyone want to comment on that?

Operator, could you check the phone line for any comments on this topic?

Operator: If you have a question or comment, please press star and the number one on your telephone keypad.

There are no questions or comments at this time.

Dr. David Berglund: OK. Thank you, operator.

Well, we can look quickly here at a couple of others. We've got idiopathic pulmonary fibrosis which generally causes a usual interstitial pneumonia histopathologic pattern and it's you know it's one of the idiopathic interstitial pneumonia at which we have a number. And I'll try to briefly go through some of those. I don't know that we'll have time to complete all of them.

Again, there's idiopathic pulmonary fibrosis, nonspecific interstitial pneumonitis, acute interstitial pneumonia, respiratory bronchiolitis-associated interstitial lung disease, lymphocytic interstitial pneumonia, cryptogenic organizing pneumonia, and desquamative interstitial pneumonia. All these are idiopathic interstitial pneumonias.

And we are proposing to have some specific codes created for these. This ITF is also called cryptogenic fibrosing alveolitis. So we are proposing at 516.3 to

create what would be instead a new subcategory for idiopathic interstitial pneumonia.

We would create a '.30' for NOS as well as idiopathic fibrosing alveolitis, the current title is '.3'. We would create specific codes 516.31 for idiopathic pulmonary fibrosis and an inclusion for cryptogenic fibrosing alveolitis.

Questions or comments on that?

Let me look at another one. We've got nonspecific interstitial pneumonitis. One of the idiopathic interstitial pneumonias again. It mainly needs to be distinguished from IPF and some of these other idiopathic interstitial pneumonias. And again, we have a request for this to be created. Each of these do have their own clinical courses.

This one, I did propose this a little differently than it was originally proposed to us as 516.32 Idiopathic nonspecific interstitial pneumonitis. And if it was nonspecific interstitial pneumonia NOS or due to unknown underlying cause, we would ask that it be coded to 516.8 based on (exclude) note here.

Any comments on that, Dr. McCormack, or anyone in the audience?

And operator, could you please check the phone lines for either IPF or nonspecific interstitial pneumonitis to – whether there's questions on these?

Operator: If you have a question, please press star and the number one on your telephone keypad.

Dr. David Berglund: Hearing no questions, I will go to Linda Holtzman here.

Linda Holtzman: It's actually a clinical question and I guess a documentation question. The term "nonspecific" here, that is part of the diagnosis. It's not just a descriptive phrase. It's actually known as "nonspecific interstitial pneumonitis".

Dr. Francis McCormack: That's correct. It is a pathologic description.

Dr. David Berglund: So that term would show up in the record. It wouldn't just be left to the coder's discretion that this must be nonspecific.

All right. I don't know if we will just lose our phone line or what here shortly. Shall I – Donna, shall I try to do another one of these or shall we break for lunch?

We will break for lunch now, and we will come back with acute interstitial pneumonia, not personally, you understand. And I appreciate all of the comments and input and I appreciate Dr. McCormack for being here.

Donna Pickett: OK. As David just said, we will break for lunch. We will return promptly at 1:30.

Operator, thank you. You can close the line.

Operator: Thank you. This concludes this portion of today's call. You may disconnect.

END