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The Clinical Significance of Imaging in the Management of ADPKD

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Dr. Lee:
Hello, and welcome to this webcast titled "*The Clinical Significance of Imaging in the Management of Patients with ADPKD*". I'm Dr. Jimmy Lee, Associate Professor of Radiology with the University of Kentucky. I'm joined today by one of the foremost experts in the study of ADPKD, Dr. Arlene Chapman. Dr. Chapman, please introduce yourself.

Dr. Chapman:
Sure. Thanks, so much, Dr. Lee. Um, it's very nice to be here with you, uh, and I am currently Professor of Medicine and Chief in the section of Nephrology at the University of Chicago. Thanks for having me.

Dr. Lee:
Before we get started, I want to acknowledge Otsuka America Pharmaceutical Incorporated for providing the independent education grant to support this presentation. And as you can see here, Dr. Chapman and I report the following financial disclosures.

OK, well let's, uh, review the learning objectives for this webcast. Upon its conclusion, participants should be able to summarize the strengths and weaknesses of diagnostic modalities to determine how to properly calculate TKD, apply a- the criteria for diagnosing ADPKD using abdominal imaging, describe the applications of diagnostic data to determine whether a patient is considered a rapid progressor, summarize the role of radiologic imaging, uh, plays in the management of patients with ADPKD.

So, uh, Dr. Chapman, I'll just go ahead and turn it over to you for an overview of the disease process.

Dr. Chapman:
Thanks, Dr. Lee. So, autosomal dominant polycystic kidney disease is often acronymed ADPKD. It is the most common inherited kidney disorder and the most common genetic cause of renal failure. It was first described more than 300 years ago in the King of Poland. Uh, and it's characterized by relentless kidney cyst growth, often leading to hypertension and ultimately renal failure. It's a multi-system disease and it is slow and progressive. And even though this is an inherited disorder, patients typically seek medical care in adulthood. There are between 1 and 500 and 1 in 1,000 individuals with ADPKD, yet only 300,000 are diagnosed in the United States. It is the fourth most common cause of renal failure, and it's responsible for 5 to 10% of patients undergoing dialysis therapy or receiving renal transplantation.

Here is a very nice image of a normal kidney in the middle and a bisected polycystic kidney on either side. You can see that the most significant manifestation of ADPKD are the presence of cysts, uh, and in the context of increases in kidney size. Some of these cysts are hemorrhagic, as you can see around the cortex, they're quite dark. The other thing you can see is that the cysts are focal in nature. Uh,

this Is not a diffuse disease, but a focal disease. So, even though this disease begins at the time of conception, disease progression in ADPKD is slow, it takes decades and decades before a clinical presence occurs.

Dr. Lee:
OK. Great. Thank you, Dr. Chapman for that wonderful overview. And I'll move into the imaging of ADPKD. Um, and as we, kind of, learned in medical school, uh, that we generally describe, um, ADPKD as autosomal dominant, uh, polycystic kidney disease or autosomal recessive, uh, polycystic kidney in two general, kind of, categories. And you can see that when we see the imaging manifestation, um, ADPKD is generally, um, associated with enlarged kidneys of varying sizes. Where, when we do see the autosomal recessive polycystic kidney disease at imaging, uh, which we rarely do, uh, just because of, um, the disease process, we see that the cysts are generally smaller, uh, and more uniform in size. Um, so imaging is essential for the diagnosis but can be very non-specific. Um, various morphologies, um, can, can, um, result from a single, um, etiology. And similar appearances from, um, different etiologies are s- occur, as well. So, it's very important, as Dr. Chapman had eluded to to identify the family history, the clinical presentation, and also look at other organ involvement. The imaging modalities that we use f- to diagnosis ADPKD, uh, generally is family history plus, um, some, sort of, imaging finding. Uh, and in most cases, that's gonna be ultrasound. Greater than 98% of at-risk individuals are identified by age 30 using ultrasound, um, as the most common modality.

When we talk about imaging follow-up, uh, we generally would like to use MRI. Um, and that's the most commonly used, just because as I previously said, we're diagnosing this patients at age 30 and MRI does not, uh, utilize ionizing radiation like CT does, um, and it serves as a novel, uh, surrogate, kind of, a biomarker for kidney disease, uh, to determine clinical endpoints for both, um, identifying the disease severity and also for clinical trials.

Um, we use a lot of other modalities to identify the complications that are associated with, uh, ADPKD, uh, such as dual-energy CT. We can help characterize some of those stones that, uh, was previously mentioned. We can also identify hemorrhage or even tumor, uh, involvement and sometimes infection, uh, when that happens to some of the complicated cysts.

Here's a nice, uh, slide, uh, that shows the different imaging modalities. In the top, left-hand corner, you can see how the typical appearance of, uh, ADPKD with ultrasound with, you have cysts that are of varying size. And, uh, mostly are ging to be anechoic. But some of them can be complicated by hemorrhage or protein. Uh, on the top right you can see a non-contrast CT and a contrast-enhanced CT. Again, you can see the multiple cysts of enlarged, uh, kidneys of, uh, with cysts of varying size. On the bottom right, you'll see a T1 weighted, uh, non-contrast enhanced MRI, uh, and the very bottom right you'll see a T2 weighted, uh, MRI that shows, um, the cyst, both in the kidneys and, uh, in the liver.

So, imaging findings of, uh, autosomal dominant polycystic kidney disease, um, there's a lot of variability like Dr. Chapman had talked about. We can see, um, these two patients here that one has, uh, the type 1 mutation in the other one to the right has the type 2 mutation. Uh, these are at similar ages, but you can see that the PKD1 mutation, the cysts are already enlarged and they, um, the kidneys are massively enlarged, as well. Uh, whereas the PKD2, uh, mutation, the kidneys are not as enlarged, uh, with, with more of a limited involvement of cysts. Furthermore, um, even within a patient, you can see that, uh, you may have unilateral disease, uh, where one kidney is affected, and the other is normal. Uh, or y- may even have focal or, uh, segmental disease involving the kidneys. So, uh, you'll see in this case example that there's only involvement of the lower pole of the right kidney. Sometimes, you'll even have, uh, genetic abnormalities that are closely related. So, here's an example of, uh, polycystic kidney disease with tuberous sclerosis. You can see that there are both cysts and fat-containing, uh, angiomyolipomas, or AMLs. Um, just to review, a few things that, uh, Dr. Chapman had said, um, most of the patients that we encounter in daily practice are gonna be, uh, PKD1 patients but we'll also see the PKD2 patients. Uh, but you can make these general ide- generalizations that if you see a younger patient with lar- enlarged kidneys, multiple cysts, um, they are probably going to be the, uh, autosomal dominant polycystic kidney disease, uh, the type 1. And if they are older, um, and they s- have smaller cysts, uh, you could a- again in general, uh, you might be able to think, uh, that they are the type 2. But again, these th- it's very heterogenous and it's really, at this point, no way to determine if you have a, uh, type 1 or a type 2, um, mutation, uh, other than having a genetic test performed.

Um, but again, you can see at the bottom of this slide, the side-by-side comparison, um, in the bottom left-hand corner, you can see a young individual with, in the teens, twenties, uh, small cysts, the kidneys are still of normal size, where in, um, when they get into the forties and fifties, they have enlarged kidneys with, um, multiple, multiple cysts. And to the right you'll see a patient that has the, uh, type 2 mutation and at age 50, uh, they, their kidneys are not as enlarged, uh, but they do have multiple cysts of varying size. And then once you get to age 79, um, they start getting to, um, the more typical pattern that we see and associate with when we think of autosomal dominant polycystic kidney disease. Here again, just to, um, illustrate that point once again with MRI, here are coronal T2 weighted images, um, side-by-side, both 42 year- 43-year-old males, uh, but you can see that phenotypic variability of the disease process here.

Uh, mosaicism, uh, just refers to, uh, the process where the patient may actually have, uh, both normal and abnormal, uh, genetic

mutations or non-mutations and you can see again what we previously described where you get the autosomal dominant polycystic kidney disease affecting just one kidney or affecting, uh, just a segment of each kidney. In this classification system, uh, to describe the phenotypic heterogeneity, uh, was developed by the Mayo Clinic, uh, where you can see that they described ADPKD as unilateral, segmental, asymmetric, they also use the word lop-sided. Um, and then sometimes they have, uh, ADPKD atypical, uh, where you have either atrophy of both kidneys or atrophy of just one kidney.

Just a quick reference to, uh, autosomal dominant polycystic liver disease, um, and contrasting that with autosomal dominant poly-, uh,-cystic kidney disease. Both have hepatic cysts. The polycystic liver disease has very few renal cysts. They will both have renal cysts but, uh, nothing compared to what we see in autosomal dominant polycystic kidney disease. We often see a lot of cysts in the liver, uh, with autosomal dominant polycystic kidney disease, but it's typically later.

Some of the other associated cysts that we Uh, identify in autosomal dominant polycystic kidney disease are pancreatic cysts that we see in 5%, uh, sometimes we also see seminal vesicle cysts in our male patients, about 40% of the time. This is not associated with ovarian polycystic disease. Here's just an example of, uh, autosomal dominant polycystic kidney disease, um, on the left on the coronal MRI and to the right you can see that there are these two seminal vesicle cysts with the arrows pointing to them, uh, and down, uh, within the pelvis.

So, Dr. Chapman, I'm gonna, um, come back to you, as far as the diagnosis of, uh, ADPKD.

Dr. Chapman:
OK. Thanks Dr. Lee. So, to start with, um, it's important to remember that ultrasound is typically our imaging modalil- modality that's used to diagnose ADPKD. It's, um, inexpensive, it's accessible to all. And, uh, cystic number, uh, really does drive the diagnosis. Um, because approximately 15% of individuals do not have a positive family history, we separate the cyst number criteria for a diagnosis of ADPKD into those with a positive family history, meaning an affected parent and those without a family history, meaning that neither parent has been diagnosed. So, for those with a family history and because PKD2 is milder than PKD1, um, one really does need to wait until the age of 40 years to definitively say with imaging that someone is unaffected with ADPKD. If someone has a family member, a parent and they're at risk for having ADPKD and they're younger than 40 years of age, uh, at least three cysts distributed bilaterally is needed to make a diagnosis. And because simple cysts increase in the general population with age, the number of cysts required to make a diagnosis in older individuals with ADPKD increases, as well. So, for those between 40 and 60 years of age, 4 cysts distributed bilaterally is required and for those over 60 years of age, at least 8 cysts distributed bilaterally is required. Now, as you've seen from all the beautiful images that have been shown to you by Dr. Lee, most of these patients have far more than this number of cysts. And so, it's usually quite straight forward to make a diagnosis. And I think something that is really important to consider here is that renal enlargement is a key and unique feature of ADPKD, even though it's not included in the diagnostic criteria.

So, because, um, there are a number of people that do not have a family history or they're adopted and they're unknown, um, when someone comes with the question of whether or not they have ADPKD, the number of cysts required to make a diagnosis in those individuals is much, much, more. So, they need to have at least 20 cysts distributed bilaterally and they need to have consistent phenotype. That consistent phenotype will include something like polycystic liver disease, uh, as was mentioned, perhaps cysts in the pancreas perhaps seminal vesicle cysts. And if it's unclear that there i- whether or not there is a consistent phenotype, this may be where imaging alone is not sufficient to make a diagnosis.

So, if we look at how these cysts number criteria work, we can look at it both as a negative predictive value or a positive predictive value. And so, you can see on the left, here, this age difference in severity shows up so that if someone does not have a cyst and they're relatively young, the chances of that person having PKD1 are less than someone with PKD2. Just because cysts show up much later in PKD2 individuals.

Similarly, if someone has cysts and they are under 40 years of age, the predictive- positive predictive value for this is very high in both groups. Uh, and it's only until someone reaches the age of 40 that there is 100% sensitivity, uh, for the presence of ADPKD.

So, how do we follow these individuals? This is a very nice graph, very similar to a previous image that you saw. And again, it's really important to note that these four individuals all have the same serum creatinine. It's all normal. It's all 0.9 mg/dL. And if we were only to follow kidney function in ADPKD, we would not be able to see the differences in cyst burden. You can see in these progressively enlarging kidneys that cyst burden is increasing more and more and more as the different individuals get older. At some point the kidney can no longer compensate and residual kidney function disappears and estimated GFR declines. So, cyst burden is increased for decades before there's any loss in kidney function in ADPKD. And a measurement of that cyst burden, meaning total kidney volume has become the most important predictive biomarker for this disorder. And it's FDA approved, uh, for clinical trial enrichment.

When we were, uh, beginning the CRISP study, which is still going on today, individuals were recruited into the study who were

relatively young, who had not yet lost much kidney function. And the population was saturated with clinical risk factors for progression to renal failure. So, two-thirds of the CRISP population needed to either have early onset hypertension or detectable proteinuria. At the end of the first three years of CRISP, kidney volumes were mapped out for each individual in this study. And there were 241 individuals who started in CRISP. And what you can see here are curvilinear growth curves. Each individual that are slightly different from each other, but their consistent within that individual. And there are some individuals on the left-hand side of the graph who have very aggressive disease and very large kidney volumes, even though they're less than 21 years of age. On the other hand, there are a number of individuals on the lower right-hand side who have relatively small kidneys but are not growing very fast, at all.

The other thing you can see is that if you take the graph of cyst volume, you can pretty much superimpose that graph over the kidney volume. And this suggests that the renal enlargement that we see in ADPKD is due solely to the increase in cyst volume. Now, we expected that, um, the PKD1 patients in CRISP would have bigger kidneys than the PKD2 patients. And in actual fact, they do, they are significant bigger, but what's fascinating is that they grow at exactly the same rate. So, we went back and counted the number of cysts in the kidneys of PKD1 and PKD2 patients and the PKD2 patients have approximately 40% fewer cysts than PKD1 patients. So, the differences in disease severity between PKD1 and PKD2 individuals is really due to cyst number, not to rate of cy- kidney growth.

Dr. Lee:
Thank you Dr. Chapman. Where my, my experience with measuring, um, total kidney volume started, so, uh, working with my nephrologist here at the University of Kentucky, uh, he originally approached me and asked, 'Can you give me, um, kidney, kidney volumes for my, uh, polycystic kidney disease patients?', and my immediate visceral reaction was, uh, probably not because that, um, segmentation was mostly manual, at that time, um, and that would add anywhere from 15 to 20 minutes to reading a study, uh, that I could otherwise, um, be done in maybe 5 to 10 minutes. Uh, so, um, it really added, you know, doubled the reading time to that. But we looked at the, uh, papers that were being published by, uh, the CRISP, uh, group, uh, Dr. Chapman's group and came to realize that we could, uh, in fact use the ellipsoid formula, uh, for clinical purposes. Um, so really, um, we already do this in radiology quite a bit and different, uh, areas, uh, when we measure kidney volumes for donors, um, we often use the volume of an ellipse when we measure volume for ovaries, we also use, um, uh, the, the formula for an ellipsoid. Um, so here you can see that we do the exact same thing for the autosomal dominant polycystic kidney disease. We do three perpendicular measurements. Uh, we choose the coronal plane for, uh, the cc measurement, the coronoal-, uh, craniocaudal dimension. uh, we go transverse and, um, anteroposterior using the axial images and you can see that, uh, we use this, uh, formula at the bolo- bottom. Uh, but most radiologists have this memorized and just have simplified the pi divided by 6 and realize that you just take all those three-dimensional measurements, and you multiply it by 0.52. Um, now if you don't wanna do that, you can go to, uh, some of these websites and you can type in, uh, some of the, um, measurements in millimeters, here and it will calculate the total kidney volume. You can also, um, and it will go ahead and sum it, as well. Uh, you can also add in the height, which will, um, normalize it for that patient.

Uh, and what I want you to see here, is that, um, it really, uh, using the Mayo classification of total kidney volume, uh, it puts people in class, uh, A through E. And what, um, I didn't realized, um, when, uh, st- I first started doing these, uh, volumetric measurements for my nephrologist was what their, um, idea was, was to identify these patients that were in class 1C, 1D, and 1E, um, as those that were rapid progressors as people that they knew that they needed to keep an eye on, clinically or even start, uh, therapeutics, um, because even though their renal function was normal, um, these people were identified as, um, at-risk individuals, um, based on their total, uh, kidney volume. So, uh, it w- really m- was eye-opening to me how, um, really just three measurements and a simple formula could add so much to the patients' care.

Now, when you start reading, um, and this is what I did initially when I was approached to do t- kidney volumes, you'll read, uh, that the CRISP study used, uh, stereology, uh, for their measurements and most of this, um, is used for research, uh, purposes, uh, at this point but, uh, as radiology grows and artificial intelligence, a lot of these, uh, applications will become more and more available to us and we'll be able to apply it in our daily work.

They also talk about, uh, planimetry. Uh, you'll see a lot of papers on planimetry and how, um, if you manually or semi-automatically, um, trace that border of the kidney, you'll be able to, uh, identify the volume, uh, with more accuracy. And here, you can just see that, um, as far as time, accuracy, and reproducibility, um, you can go from left to right. Where the ellipsoid measurements have the lease amount of time commitment, uh, but also will have the least amount of rep- the, the worst performance, as far as reproducibility and accuracy. And when you go over from stereology into planimetry, um, you get more and more accurate. Uh, more and more reproducible. But also, you'll notice that, uh, the time commitment goes, uh, goes much higher.

Um, here you can just, um, these are some slides that were provided by our colleagues at the Mayo Clinic. Uh, they have automated total kidn- kidney volume measurements that they're developing. Uh, and this hopefully will be available in a broader sense, not just in research cen-, uh, centers where we'll be able to apply this in everyday clinical practice.

As far as complications for ADPKD, there are a lot of things that we, uh, definitely are on the lookout for. Um, often patients will present with pain, hematuria and fever and typically, um, those are associated with, that's a sign that there may be, uh, cyst hemorrhage. Uh, as you can see on this ultrasound, um, the red arrows indicate an area of, uh, l- likely hemorrhagic, uh, conversion of a simple cyst. Uh, here again, you can see the hyperattenuating area surrounding a cyst, uh, that suggests that there has been hemorrhage. And a lot of times you'll need serial examination. So, you'll be comparing the patients prior exam to the current exam when they're presenting with pain, and you'll use that to identify whether the patient, uh, whether we can find or narrow down the reason for the patient's presentation.

Here again, where you can see a cyst actually ruptured, the area of increased attenuation indicated by the red arrow, uh, shows or suggests that there's hemorrhage. You can also see that there's asymmetric peri-nephric, uh, fat stranding or edema, um, that also indicates that that cyst has undergone either hemorrhage or some other complication.

Another complication that Dr. Chapman had mentioned was stone formation. Um, later on in the disease process, we see stone formation up to, uh, and up to 20% of those patients and a lot of times, they are uric acid stones, about 50% of the time. And that's where dual energy CT can, um, help us identify, uh, uric acid versus non-uric acid stones. Um, and, uh, as you can see in this, uh, coronal reformatted image, here.

Another common, uh, process that we see, or complication is infection with autosomal dominant polycystic kidney disease. Uh, sometimes we see this cyst have air-fluid levels, which would suggest a gas forming organisms. Uh, some of the times the cysts have peri-nephric edema. Um, a- so a lot of times it will mimic a cyst hemorrhage and it's really the clinical, uh, presentation that allows us to differentiate between the two.

Um, another complication, um, although, uh, ADPKD is not associated with an increased risk factor, risk for renal cell carcinoma, it doesn't mean that they do not have renal cell carcinomas. And the challenge, really with ADPKD patients is that their kidneys are so complicated. So, they have many cysts with m- many varying attenuations on CT or signal intensities on MRI and your job, as the radiologists is to determine which one, um, could potentially have neoplasm within it.

You can see, uh, we have an ultrasound example here where there is a complicated cyst with vascularity i- in area of soft tissue, uh, echogenicity, uh, shown by the colored Doppler imaging. Here on the coronal MRI, you can also see that there's, uh, soft tissue within the cyst wall on th- in this right kidney. And if you give contrast, you can see that cyst wall enhances with a mural nodule. Here is an additional patient on the left kidney. Uh, you can see again there's a distinct cyst with soft tissue instead of, uh, cyst fluid or hemorrhage. Occasionally, uh, you'll use your diffusion weighed imaging which can really help distinguish between those just hemorrhagic or complicated cysts versus those who may have, uh, converted into renal cell carcinoma.

So, with that I'm gonna go to Dr. Chapman, um, on the treatment of ADPKD.

Dr. Chapman:
Thanks, Dr. Lee. So, I'm going to talk to you starting with a case presentation and we're to use some of the imaging tools that Dr. Lee just reviewed so nicely with you. So, we're gonna talk about a 38-year-old woman. She has a normal serum creatinine of 0.9 mg/dl, which translates to an estimated GFR of 81 ml/min. She has a height TKV measured, and we correct TKV to height, um, when we do the Mayo classification so that she has a height TKV of 1,987. Now as I showed you before, there's a curvilinear growth curve that's unique to each individual and it's constant, so we can use this one data point, one point in time and determine how fast these kidneys are growing, assuming that at the beginning of life, patients had a normal kidney size.

So, for this young lady, uh, at the age of 38, you can see on the Mayo classification, her height-corrected TKV is shown with a dot, and she sits in the most severe classification, 1E and it's highlighted with an arrow. So, it's not surprising that she does have a PKD1 mutation and her PKD1 mutation is a truncating mutation, the most severe form of mutation that you can identify. So, she sits in a high-risk, rapidly-progressing patient with ADPKD. So, we can take this patient and we can look at her annualized growth rate and if someone is a 1E patient, uh, they grow more than 5.5% per year, which is equivalent to about 80 to 90 ml per year, which is equivalent to about half a kidney per year. So, they're growing very rapidly. And if you can see in this individual that when she started tolvaptan therapy, um, there was an immediate sharp decline in the rate of her total kidney volume growth. And then as time went on, uh, she came back into a growth rate, but a much reduced growth rate, at 4% per year, again at 4% per year, and then slightly less at about 3.5% per year. So, this is one way to use total kidney volume as an estimate for effectiveness of therapy when one looks at the rate of kidney volume growth per year.

So, um, tolvaptan was approved for patients with rapidly-progressive disease in April of 2018. And based on a number of molecular pathways, there are a number of different drugs either being tested pre-clinically in animal models or in humans in phase 2 or 3 clinical trials. Uh, as I mentioned, the vasopressin V2 receptor antagonist, tolvaptan, was tested in the TEMPO trials. There have been tyrosine

kinase inhibitors that have been tested. Uh, there have been somatostatin analogues, uh, and statins that, uh, activate GS proteins that have been tested. There are mTOR inhibitors, uh, targeting, um, cell cycle regulation. And there have been drugs that block the cystic fibrosis transduction channel, as well as potassium channels, uh, known to inhibit, uh, fluid secretion into cysts. Uh, more recently there has been, uh, uh, activity in clinical trials looking at bardoxolone methyl, as well.

Dr. Lee:
So, thanks again for all the, the great insight. Um, I'm just gonna sum up our talk, uh, with a few take-away, uh, key take-away points.

So, um, autosomal dominant polycystic kidney disease has an incredibly wide range of phenotypic variability. I think, um, we've illustrated a lot of the, uh, the variability that we encounter on imaging. And, uh, but Dr. Chapman and I, um, spoke to the measurement of TKV as being the best biomarker for staging and predicting disease progression, currently. And really, um, the reason that we're here together is to emphasize the importance of a strong, uh, radiologist/nephrologist reletio- relationship, um, providing the necessary information to aid in patient care.

So, with that, uh, Dr. Chapman, I'd like to invite you to describe some of the, um, relationships that you've built at your own institution with radiologists and how those interactions have gone.

Dr. Chapman:
Sure. So, um, you know, I think there, that it's really an important relationship, particularly in polycystic kidney disease. Um, you know, we've worked hard with our, uh, abdominal imaging specialists in the department of radiology to really try to refine the information that we get at the time of, uh, diagnostic imaging. Um, and that being what we really have been able to fortunately be able to get is a pretty consistent report where, uh, we get a total kidney volume measurement. Um, we can do it ourselves, uh, on our PACS system but I think it's always much better when a radiologist does it 'cause that's what they have the experience and the skill to be able to do. So, that's been one key relationship, um, that w- we've developed.

Um, the prognostic issues, I think, fall more in my court to get back to the radiologist about, but the radiologists are now going to the Mayo classification and plotting the data and actually providing an estimate of whether or not a patient is a 1A, B, C, D, or E. Um, they are also very helpful at telling us th- if it's atypical PKD that we really shouldn't be using the Mayo classification, at all.

Dr. Lee:
Right.

Dr. Chapman:
Um, s- so that's also been very, very helpful.

Um, the treatment in ADPKD, um, as you saw on the case and also based on the rate that these kidneys grow, um, we don't image that frequently, um, usually at the most every other year. Um, but when we do, we do reach out to the radiologists and ask if they could possibly do a comparison measurement, just to see. And then obviously for all the complications, we, we rely heavily on our radiologists to help us, you know, to see this stone, see the hemorrhage, tell us whether or not they think that there's an air-fluid level or if there's a suspicious mass in the kidneys. So, yeah, it's a really important ongoing collaboration and partnership.

Dr. Lee:
Right. And, uh, you know we shared some similar, uh, interactions here at the University of Kentucky. I think I mentioned, uh, earlier, uh, when the nephrologist asked me for the first time to do total kidney volume, uh, I was a, a bit hesitant, but after reviewing some of the literature and really, um, working with him, I understood the importance of it. So, you know, that was very good of him to really take the time to, um, explain to me the importance of providing that measurement in patient care.

And one of the things that we were able to do, um, just because we were starting it from the ground up and any radiologist that are interested in doing this and starting it at their own program, is that we built an MR imaging protocol that was very abbreviated. Um, and, and followed the, uh, recommendations that were outlined in some of the, uh, the CRISP papers that h- h-, um, have come out, where we really just did, um, a non-contrast MR of the abdomen, um, and again w-, you know, the, t- for a radiologist, it's, it's almost, before we used to fear the (laughter) the autosomal dominant polycystic kidney disease because we knew we had to look at every single one of those cysts and try to identify any complications, and we still do that. Uh, but, um, we didn't find those complications too often. But to have that added value of being able to provide something that you can use as a nephrologist to really make a decision upon that patient can really give us more insight into how we are contributing to the, to the care of that patient.

Um, and again, what you were talking about, um, standardized reporting, um, knowing that when we see an autosomal dominant polycystic kidney disease patient, that that's something that is automatic now, that we're gonna give you a, um, a total kidney, uh, volume so that you can make that decision and not have to d-, you know, take time out of your busy day to ask for, um, just a simple

measurement. So, um, that's something that if again working together closely with your, um, nephrologist/radiologist relationship, developing that, um, both imaging protocol and the report is, is very important.

Um, one question, um, I have for you, Dr. Chapman, as far as, um, do you eve encounter where the radiologists may be the first person who discovers the disease process? And, um, then you go back and maybe, you know, the patient's family actually did have kidney disease but maybe was never diagnosed with autosomal dominant polycystic kidney disease?

Dr. Chapman:
Yeah, that absolutely does happen. Um, you know, we even actually had people who've been evaluated for other medical problems happen to be diagnosed at the time. So, usually there's some reason that they needed to get imaged.

Um, I can think of one recent case of a gentleman who had a, um, descending, uh, aortic dissection, uh, and they imaged the abdomen because it continued down to the renal artery and they discovered polycystic kidney disease, and he actually did have a family history.

Um, another good example is someone who had been followed for many years, um, for sarcoid and was actually getting ready to go lung transplantation and they discovered polycystic kidney disease. So, you know, you can find it, but something has to trigger the need for some imaging.

Um, one area that I was hoping could potentially develop further in radiology is this idea of, um, a rapid scan. You know, I think it's done for other, uh, type of lesions, like pancreatic cysts, where patients need follow-up, and they get a very brief image done. Um, and if we were able to have something like that, as you were saying you do at your institution where you get it done in a very short period of time-

Dr. Lee:
Right.

Dr. Chapman:
-uh, we would probably image people sooner on therapy-

Dr. Lee:
Right.

Dr. Chapman:
-than we do right now. Um, I mean it's somewhat cost-prohibitive-

Dr. Lee:
Right.

Dr. Chapman:
-to do the full MR of the abdomen and so I think that's part of the reason we wait that two year interval to think about imaging.

Dr. Lee:
Right. And you're, you're touching on something that, uh, is, kind of, beyond my expertise but, and, and, there is a push for, um, a abbreviated MRI, um, billing code, um, where-

Dr. Chapman:
Wow.

Dr. Lee:
-we hope that that will, you know, we're doing most MRIs of the abdomen take anywhere from 30 to 45 minutes on the table, but-

Dr. Chapman:
Mmmhmm.

Dr. Lee:
-the abbreviated MRI, the hope is that we, um, can use this in multiple different areas like you were talking about, the rapid. But, um, charge the patient less, because it really does take less time, uh, both MR imaging, um, and with interpretation, as well. So, I think that is definitely something in the future and we're hoping that, um, some different billing codes will come out of MRI where we can, we can utilize that abbreviated MRI is, kind of, the buzzword that you should be looking out for, hopefully.

Dr. Chapman:
Wonderful.

Dr. Lee:

So, Dr. Chapman, thank you again for, uh, being with us here today. I've really enjoyed our time together. I've learned quite a bit from you, um, and this concludes our learning activity today and thank you, uh, to all the participants.

Dr. Chapman:
Absolutely, and thanks for having me.

Announcer:
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