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## ***Advances in PH Journal***

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"//connect.facebook.net/en_US/all.js#xfbml=1"; fjs.parentNode.insertBefore(js, fjs);
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Vol 10, No 3 (Fall 2011)

## The Challenges of PAH in Pregnancy

PAH patients who become pregnant—certainly against medical advice— or whose disease is diagnosed while pregnant present myriad challenges. On October 17, 2011, a group of physicians who have dealt with these complicated issues met by telephone to discuss their approaches and thoughts on dealing with these unique patients. Co-guest editor **Deborah Jo Levine** facilitated the discussion among Drs **Ron Oudiz**, Director of the Pulmonary Hypertension Center at Los Angeles County Harbor-UCLA Medical Center; **Irene Lang**, cardiologist at the Medical University of Vienna; **Evelyn Horn**, Director of the Heart Failure, Mechanical Circulatory Support, Pulmonary Hypertension Program at Cornell and adjunct

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professor at Columbia; and **Dianne Zwicke**, Medical Director of the Pulmonary Hypertension Clinic at Aurora St. Luke's Medical Center in Milwaukee, University of Wisconsin School of Medicine.

### **Dr Levine:**

Good morning, this is Debbie Levine and I would like to thank everybody for joining me and participating in this roundtable discussion. This issue of *Advances in Pulmonary Hypertension* is devoted to several topics concerning women with PAH. We hope to devote the next hour together to discussing an issue that, although not truly very common, is very complicated and poorly defined. We will discuss the evaluation, management, and therapy of the pregnant patient with PAH. Most often, these are women who are initially diagnosed with PAH during their pregnancy, and occasionally a woman with known PAH who becomes pregnant against medical advice. In either case, we'll discuss the options available to optimize the chance of successful maternal and fetal outcomes for those women who opt to carry their child to term. The true incidence of PAH in pregnancy has not recently been reported, but there are multiple case studies over the last several years showing a mortality rate ranging from 25 to 60%. The more recent studies that report mortality as low as 25% or less show some improvement in outcomes. But despite this apparent improvement, the risk of maternal death is still unacceptably high. Patients with PAH should always be advised to avoid pregnancy. Contraception and counseling for this first-line recommendation are discussed and detailed within other sections of this issue of *Advances*, so here we will focus on the approach each one of us takes when we encounter these patients in our own centers. Before we get started, can everyone mention who they are and where their center is.

### **Dr Horn:**

I am Director of the Heart Failure, Mechanical Circulatory Support, Pulmonary Hypertension Program at Cornell. I am an adjunct professor at Columbia and I continue to advise my Columbia colleagues for much of the high-risk cardiac OB program, particularly these patients.

### **Dr Lang:**

I am Irene Lang from the Medical University of Vienna, Austria, and I'm a cardiologist working in the interventional lab and I have a clinic for pulmonary hypertension together with 2 other physicians.

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**Dr Oudiz:**

I'm Ron Oudiz, a cardiologist at the Los Angeles County Harbor-UCLA Medical Center, and the Director of the Pulmonary Hypertension Center here, working with 2 of the most excellent PH nurses.

**Dr Zwicke:**

Dianne Zwicke. I'm a cardiologist and the Medical Director of the Pulmonary Hypertension Clinic at Aurora St. Luke's Medical Center in Milwaukee, Wisconsin. We are academically affiliated with the University of Wisconsin School of Medicine and Public Health. I also cross-cover the mechanical assist device, advanced heart failure, and heart transplant services.

**Dr Levine:**

Great, so glad you all could attend today. Why don't we start with the type of early approach we take when a patient is first referred to us—either a pregnant patient newly diagnosed with PAH or one of our patients with known PAH who becomes pregnant? What approach do you take, in terms of getting the right people in your center together to help to evaluate, monitor, and manage these patients? Evelyn, can you tell us how you start off with that?

**Dr Horn:**

We should first address the possibility of early termination, depending on when during pregnancy they are presenting.

**Dr Levine:**

Okay, this is a good place to start, can you tell us how you go about getting to this point?

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**Dr Horn:**

The first aspect is for any woman who has not had a right heart cath. Again, everything that we are talking about, particularly termination and mortality, hinges on the definitive diagnosis of PAH; this means having right heart catheterization data even during pregnancy. If a woman comes to me from another institution with an echocardiographic diagnosis of pulmonary hypertension, I will perform a right heart cardiac catheterization to confirm the diagnosis. The right heart catheterization should first be tried without fluoroscopy; and if necessary, then fluoroscopy needs to be done. I would preface all comments by saying we must be 100% sure the diagnosis is precapillary PAH and not pulmonary venous hypertension, obesity, or obstructive sleep apnea with PH. This information is critical before counseling the patient about possible termination of pregnancy.

**Dr Lang:**

I think that sometimes in the literature, these success stories cases may not be true PAH; that is, the hemodynamic diagnoses are not correct. For example, there are some pregnancies that are described being managed with very little medication who make it through until the last month of pregnancy. And sometimes I'm not sure whether pulmonary hypertension is truly precapillary because they have this large mixture of postcapillary patients who look just like precapillary. I think what you said is very correct. You need to do a good right heart cath and be sure about the pulmonary capillary wedge pressure (PCWP). If there is any question about the accuracy of the PCWP, then one should obtain a left ventricular end diastolic pressure (LVEDP) in parallel just to make sure that you're looking at precapillary disease.

**Dr Horn:**

Right, and emphasize, don't be afraid to do that catheterization even while they're pregnant.

**Dr Lang:**

Right, exactly.

**Dr Horn:**

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I think the next aspect is up until when would we perform a termination of pregnancy. In general, if a woman presents early, and once that diagnosis is made, much of the focus is spent on termination, because I still believe that despite all of our successes with medication and differences in mortality, mortality remains extremely high and we should advise the patient to terminate the pregnancy. The question is at what stage is delivery of a dead fetus as problematic as an attempt of a live delivery? I have had a high-risk PAH patient with systemic level PAP terminate as late as 22 weeks' gestation.

**Dr Levine:**

I think this is important, as we know that a termination procedure itself is not entirely without risks as well.

**Dr Horn:**

Absolutely true. And for that procedure, one needs the same support that we would need for delivery in terms of a multidisciplinary team, which includes cardiac anesthesia, OB anesthesia, and often cardiac surgery, not uncommonly, for the sickest patients. I have actually had femoral lines placed on occasion so that if we needed to crash onto ECMO we can. Obviously, maternal and fetal medicine (MFM) needs to be present; the difference being the involvement of neonatology for live births. But I think the first aspect is to get people together—including the partner/spouse—to make sure that everybody understands the risks. I think it's very important for the spouse to understand that the worst possible scenario is that of a marked premature delivery with possible maternal death and marked disability of the child due to prematurity. So that is why lots of early efforts are made in this regard for the sickest patients.

**Dr Levine:**

So it sounds like termination early-on is an option, but how does everyone feel about timing as it becomes more of an issue as the pregnancy progresses? So in terms of being too late, is there a “too late” situation where the procedure would be as risky as the delivery itself to the patient?

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**Dr Lang:**

My obstetrician teaches me it depends on the exact stage of the pregnancy. As the child becomes bigger it becomes a delivery and it carries exactly the same risk as getting the baby. And then early-on it may be a smaller intervention, but still an intervention.

**Dr Zwicke:**

I would agree with that because by the time they're reaching 16 weeks' gestation they hormonally have had a significant increase in intravascular volume and have had a redistribution of fluid into the third space.

**Dr Horn:**

Although 16 weeks' shows significant hormonal and hemodynamic changes, we haven't seen the maximal hemodynamic changes in terms of cardiac output.

**Dr Zwicke:**

You must pay meticulous attention to the volume status of the mother and be on top of it. You need to treat that and manage it throughout pregnancy and after delivery, be it a live birth or termination. There will be different challenges depending on what the etiology of the pulmonary hypertension is.

**Dr Levine:**

So it sounds like from the discussion that the option of early safe termination may be the first discussion to have with the patient who presents early enough in pregnancy. Importantly, both for this procedure as well as with continuing on with the pregnancy, the next step should be setting up a multidisciplinary team of all of those involved. This will help to optimize care through this procedure or through the pregnancy and the delivery. For those patients in whom termination is not an option, for example, those who present too late in pregnancy and they are going to go through with the pregnancy—can we talk about what people's diagnostic and management algorithms are?



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**Dr Zwicke:**

So, the person who has diagnosed the pulmonary arterial hypertension is responsible for the first discussion with the patient. This discussion needs to address the new diagnosis, the implications, the effects of pregnancy, and the forward options. Within the next 3 days, it is ideal to have a joint appointment with the patient, significant other, the high risk OB physician, and yourself. The issues of termination versus continuation of the pregnancy need to be fully discussed. If she decides to proceed, we set up an entire team. We pull together the high risk OB team, neonatology, cardiology, pulmonary, and cardiovascular anesthesia, and the OB and ICU nursing staff representatives. We then get a plan in place and designate who is going to be the leader of this team, which is usually the person managing the hemodynamics, treating the PAH, and monitoring the RV function. In my opinion, it is usually the RV function that will dictate the outcome of the patient. Identification of the patient-specific issues early on makes for a safer and more efficient team, should there be an unexpected premature delivery.

**Dr Lang:**

We also got advice from a rheumatologist in my last case. She had scleroderma with antiplatelet antibodies, erythrocyte antibodies, and we thought about plasmapheresis; so in addition to the experts you just numbered, we had a rheumatologist on board.

**Dr Horn:**

I think we would all echo the same approach in terms of the multidisciplinary team. Occasionally there is a hematologist as well. But pretty much, that's most of the team.

**Dr Zwicke:**

Someone has to be declared the leader of the team, as information from any other person on the multidisciplinary team needs to feed into someone. In my experience, that's the most important single item in the planning process. Most of the issues through the 2nd and 3rd trimester will be related to right ventricular function as the most important. And if right ventricular function is not the most important, how the mother tolerates the physiological

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stressors of the pregnancy may be the most important. Do you add diuretics? Do you hospitalize? Do you deliver earlier than planned? Do you put them on dobutamine? Do you alter your initial plan in any way?

### **Dr Oudiz:**

You bring up the point of not only having a cohesive team with good leadership, but also having a multiple contingency plan. Because you don't know what's going to happen nor when it will happen.

### **Dr Zwicke:**

Well, you have to be prepared to deliver at any point in time if the right ventricle starts to deteriorate and you can't rescue it. If they ate at a Chinese buffet and had tons of soy sauce 2 days ago, you know the cause of the hypervolemia and can diurese them a little, feeling confident that you have the situation under control. Deterioration of the RV function without an identifiable cause always makes us more nervous. The beauty of the advances in echo technology is that we can reassess the right heart in an office study. We have the luxury to look at the RV function serially. I normally will look at RV function by echo every 2-3 weeks from the first time I meet the patient, moving to every week during the last 6-8 weeks leading up to the designated delivery time. A limited right heart echo only takes about 20 minutes and provides a wealth of information: RV size and function with tricuspid annular plane systolic excursion (TAPSE) with at least 4 views, width of IVC and respiratory variability, RA size, estimated RVSP, degree of tricuspid regurgitation, and RA pressure. Once the patient tells me they're short of breath or they have swollen feet; we have more objective data serially to make our treatment decisions. You really do need to know that information earlier and I believe serial echo evaluation of RV performance may help the patient assessment prior to the development of clinical symptoms.

### **Dr Horn:**

We usually have multiple contingencies in terms of planned delivery, emergency delivery; including where, how, what before and after delivery. Are they on the cardiology service? Are they in the CCU or—depending on the comfort level of the nursing on labor and delivery—are they on a high risk L & D unit? Is delivery in the cardiac OR versus on labor and delivery?. Is OB nursing going to cardiology or is cardiology nursing going to L & D? All of these things have to be addressed. And we should also address the patients who are new to us who just come via our emergency room, are transferred with this diagnosis previously not on therapy,

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or patients who are cared for elsewhere already on therapy. I think all of these things vary from case to case. Does the patient have a shunt? Is this congenital heart disease which makes it a little bit more favorable in terms of filling of the left side or is it a patient who has all the bad prognostic aspects and introducing a cardiac surgeon and discussing emergency procedures if necessary is required?

### **Dr Levine:**

It sounds like evaluation and monitoring—serial monitoring by many specialties—occurs in all of our centers. In terms of a plan and monitoring, at what point and with what types of medications will you follow these patients? And at what point do you consider changing someone from an oral PDE-5 inhibitor to a prostacyclin?

### **Dr Oudiz:**

Whether we should be treating patients empirically or expectantly is an important discussion point. In other words, starting a prostacyclin when you might ordinarily otherwise not start a prostacyclin, or treating them on an as-needed basis. And the worry that all of us have is that, like Dianne was saying, if we wait until the patient is symptomatic, then it's probably too late. The problem is that there just isn't much evidence in the literature or even anecdotal experience because there is so much heterogeneity in the approach. But I think most of us at the level of a PH specialty center are aware of what the risks and benefits of prostacyclin infusion are for a patient who has an otherwise uncomplicated pregnancy. And therefore we would probably edge toward the side of earlier initiation of prostacyclin, if they're not already receiving it, rather than expectant initiation of prostacyclin.

### **Dr Zwicke:**

I would completely agree with you, Ron. The typical pregnant patient I see enter my practice is in that mid-teens' weeks of pregnancy. So, they are usually around the 16th–18th week when there is a large fluid shift occurring. They come in because of right heart failure symptoms. When you look at the echo, you see that the RA and RV are dilated and the RV function is depressed, the pressures are high, the PA is dilated, and the IVC is more dilated than what you'd expect to see in pregnancy. It is very helpful to add a bubble study (injection of agitated saline) at the time of the first echo study in your office to rule out an intracardiac right to left or bidirectional shunt – the most common congenital systemic to pulmonary shunt missed in adults is a sinus venosus atrial septal defect in an echo-silent area seen by transthoracic echo or a PFO that has opened from the elevated right heart pressures. The

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right heart catheterization is necessary to confirm your diagnosis of pulmonary arterial hypertension and allows a complete oxygen saturation run. The saturation run would also detect anomalous pulmonary veins. If a shunt is not apparent on 2-D echo imaging and you don't do a bubble study, which can safely be performed on a pregnant woman in the office, you will never know that you are dealing with a shunt and will diagnose this patient with idiopathic PAH instead. The hemodynamic measurements will definitely help you make the best pharmacologic selections for each patient. If the cardiac output/cardiac index is low or even normal, prostacyclins are definitely indicated. All pregnant women would be expected to have elevated cardiac indices. Since these patients are well into their pregnancies, you need to start aggressively treating, as delivery could occur at any time. This is a time when the "simpler" treatments are probably not appropriate.

**Dr Horn:**

And, I would like to emphasize that the normal pregnancy should have a high cardiac output, so finding a low or even "normal" cardiac output in pregnancy is markedly abnormal.

**Dr Lang:**

I would go as far as to say if it's truly PAH and the person is pregnant, I don't see their living through pregnancy without a prostacyclin. I think it's really the worst if a scleroderma patient who is pregnant gets PAH or a scleroderma patient with PAH gets pregnant. All the pregnant women I have seen have been put on IV epoprostenol.

**Dr Horn:**

I think there may be an occasional patient, for example with a large congenital systemic to pulmonary shunt, who we may not start on intravenous prostanoid therapy.

**Dr Lang:**

Right. Well I'm talking about PAH associated with an elevated PVR.

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**Dr Zwicke:**

All of my patients have been treated with IV epoprostenol or IV treprostinil. One patient, long ago, came to me on subcutaneous treprostinil. We need to remember that 20% of the cardiac output goes to the skin in a healthy person, but this can drop as low as 5% with poor RV function. Therefore, it's risky to administer a drug subcutaneously and not know what amount of drug will be absorbed from day to day, depending on the RV function. The optimal delivery system is IV.

**Dr Oudiz:**

Let me throw a little bit of a wrench into this: when you're at a public hospital and you have patients that are uninsured and undocumented you are probably okay in the inpatient setting if your drug is on formulary. And you can probably be okay with the emergency insurance during pregnancy. But for the longer term, if the patient has advanced PH and becomes "dependent" on an IV prostacyclin, you're in big trouble because you're going to have to either have the hospital pay for it or the patient pay for it.

**Dr Zwicke:**

Actually, there are patient assistance programs for several of the medications now that may apply.

**Dr Lang:**

But Ron, haven't you seen them actually get worse after the pregnancy is over? They're even worse than before so it's foreseeable that they will not be treatable just with a phosphodiesterase inhibitor after the delivery.

**Dr Oudiz:**

Oh, absolutely. That's the conundrum. We're in a situation—it's not an ideal situation—in which you can't do what you want to do, but rather have to do what you're able to do.

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**Dr Lang:**

You know I was advising in Hungary for a patient, very similar, very ill patient, and they went to the newspapers after the pregnancy was well over on epoprostenol and then she got the money from the state because it was so dramatic—a young woman—and they managed to overcome this because in Hungary there is no epoprostenol. They have iloprost inhaled.

**Dr Zwicke:**

Which doesn't work that well because of the erratic levels.

**Dr Lang:**

No, it does not.

**Dr Oudiz:**

We had a case several years ago, before we had assistance programs, that wound up being a gigantic ethical issue whereby the referring hospital ended up having to pay for the entire cost of the epoprostenol because they couldn't ethically stop it.

**Dr Levine:**

We have had the same issue in a couple of cases. In one instance, we started a patient on epoprostenol who had suboptimal funding and we needed to keep her in the hospital for a month after the delivery until she could get emergency Medicaid. She was then able to continue on with epoprostenol at home and eventually switched to treprostinil long term. So this seems as though it is not an infrequent issue.

While we are discussing medications during pregnancy, let's look into the issue of these patients being at a higher risk for thromboses. What are your thoughts on thromboprophylaxis

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during pregnancy in these patients?

**Dr Zwicke:**

If they're collagen vascular or primary etiologies, I do anticoagulate them.

**Dr Oudiz:**

What about in the beginning of the pregnancy?

**Dr Zwicke:**

Subcutaneous heparin, if we are seeing them early in the first 12 weeks.

**Dr Oudiz:**

Do you use subcutaneous heparin because of the risk of teratogenicity with warfarin?

**Dr Zwicke:**

Yes. There are good pregnancy data from England for the use of Coumadin, especially with artificial heart valves. I think they've got the largest databank available and have shown that it's safe to use the subcutaneous heparin early on, and then after first trimester, going to Coumadin, with a return to subcutaneous approaching delivery. They obviously need close observation and monitoring. If you're uncomfortable with the hemodynamics or overall condition of the patient, it is probably better to use subcutaneous the entire time.

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**Dr Lang:**

I had a patient get pregnant on acenocoumarol. We knew she had PAH, and she had a baby with a cleft-lip, a diaphragmatic hernia, and a ventricular septal defect. It was terrible. And I think the perinatal team should look at the babies early on if the patient has been on a vitamin K antagonist, because I don't think it's so rare—maybe because I have seen it.

**Dr Oudiz:**

We had an experience where we had a patient on subcutaneous treprostinil and were planning an elective delivery for her. She unfortunately went into preterm labor and ended up having to have a C-section and subsequent rectus abdominis hematoma with considerable bleeding. She survived, but we realized that the physical and temporal proximity of her subcutaneous injections likely contributed to that bleeding.

**Dr Levine:**

Thank you, these are such important anecdotes. And as we discuss delivery, we all know that we can never really plan for it. If we can, let's focus on the delivery itself. For example, when do you admit a patient for delivery? What type of delivery do you and your team feel is best? What is the best timing for delivery (if undergoing a planned delivery)?

**Dr Oudiz:**

The question is on the mode of delivery, in other words vaginal delivery versus C-section?

**Dr Levine:**

That's a good place to start.



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**Dr Oudiz:**

Well, again, I think there isn't a great amount of literature that proves one is better than the other. There are certainly those that have had more experience with one versus the other. One argument is that the C-section can be controlled in the OR and there's less variability, but that the invasiveness of the C-section and the anesthesia required might outweigh the risks of the vaginal delivery that would likely be done in an ICU.

**Dr Horn:**

And, in fact, usually the hemodynamic changes following delivery are more gradual with a vaginal delivery. Having said that, you may have to also go with your center because it takes a lot more coordination. I absolutely prefer to do vaginal deliveries and get everybody coordinated. It also depends on what week we're talking about, likelihood of favorable cervix, etc. The last thing we want to do is sort of pretend that the patient is going to go vaginally and then to crash on to a C-section. Again, with this concept if we really have things going well and we are delivering in a cardiac OR and have people hanging out waiting for 12 to 14 hours, nobody is pleased with us. But that certainly has worked. But I'm certainly cognizant of recognizing a system that can only work with a scheduled C-section. I think all of those things have to be factored in. It also makes a big difference to have the best of the team available for delivery. I should also add that another agent that I often like to use during delivery—in addition to any parenteral prostacyclin—is old-fashioned inhaled epoprostenol. It is useful as additional therapy because you can get much higher pulmonary doses. I find it even better than iloprost or nitric oxide. So it's not uncommon that I will have patients on combined inhaled epoprostenol at the highest doses tolerated and iNO. The most important issue is to be working with the anesthesiologist and to be there to make the calls for therapy for right heart, for pressors. Not uncommonly we use vasopressin as the pressor of choice and often it can be a minute-by-minute or second-by-second intervention.

**Dr Levine:**

During the C-section it is so important for us to be there with the anesthesiologist and the surgeon or the OB to help direct fluids and medications. Our input can make such a difference.

**Dr Horn:**

Yes, I believe that unless it's an emergency and we can't be there, we attend all deliveries.

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## **Dr Oudiz:**

That presents one of the biggest problems for us. That is, you never know when this patient is going to need you. And while we're on call for our patients, we're doubly on call for a delivery and really have to make ourselves available for an extended period in order to be sure that we have the best chance of actually being present. It is important that we call the shots: which pressors are used, the adjustments of the PH medications. It's very important to have the PAH expert there.

## **Dr Lang:**

What I want to point out, what I think is very important to think about, is analgesia. It's important at any time, even with the small things like an intra-arterial line. They must not have any sensation of pain during these small interventions even in preparation for a C-section or vaginal delivery.

## **Dr Zwicke:**

I wanted to share the protocol that I've used over the past decade, now for 57 patients. We assemble our team as soon as possible. We select the date for delivery early on, which will always be early in the week (Monday or Tuesday) of the 36th week of gestation. It is important to have accurate dating of the pregnancy by ultrasound. A 36-week baby is usually mature enough and none of them ended up on a ventilator. Only younger babies, some of those less than 36 weeks, required mechanical ventilation. During the last 4 weeks of pregnancy, a PAH mom with a dysfunctional right ventricle retains much more fluid due to hormonally-driven factors than what the normal pregnant mom experiences. If the patient makes it to the designated day of delivery, in the early 36th week, they come in at 5 in the morning for induction. Pitocin is used. An epidural is mandated and was discussed with the patient early on in the initial visits. It's not presented to them as an option. Cardiac and OB anesthesia work together on that so that it's a slow onset epidural, avoiding the usual hypotension and subsequent rescue with large bolus infusions of normal saline. It is important to balance vasoconstrictor medications with some saline. The goal is to have a vaginal delivery, unless there is a recognized obstetrical indication for a C-section. Most of our patients delivered by 3 or 4 in the afternoon while the team is available. The only C-sections that we have done were those with a true obstetrical reason for C-sections. One was a footling breech; another was a breech; another was transverse lie; one was very unstable with difficult to control paroxysmal atrial fibrillation; and another was a placenta previa. The interesting observation we've made out of our population is that every C-section incision

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became infected. They all had wound debridements and vacuum-assisted closure. The reason was not clear and the deliveries were performed at several hospitals, so it was not just a single hospital technique issue. That was also the information given back to me from distant centers. The vaginal deliveries do much better. With the C-section deliveries you have a much harder time judging how much fluid to take off in forcing diuresis early on day 1-2 postpartum. You usually have to wait until about the 2nd day before you can start actively diuresing them versus the vaginal delivery when we start aggressive diuresis looking at 3 L/day negative output, for 3 days, starting as soon as the epidural has worn off. That's usually as soon as they can feel their toes and wiggle them. Approximately 1/2 of the deliveries were performed here in Milwaukee, while the other half were done in other countries and other states. We actually have the fortunate situation of having a delivery room right in our ICU. There is a little foyer area for the baby, a nursery, and they can deliver right there. The anesthesiologists have to be on board with counting and estimating every cc in, every cc out, because whatever they gave during the delivery has to be diuresed off the patient. The greatest risk time for the mom is the delivery and early the morning of day 3 post partum.

### **Dr Horn:**

I would pretty much echo a very similar approach. I would also caution that we've seen difficulties at the time, and particularly with the C-section, with placental delivery. The experience is overwhelmingly much more positive from the acute hemodynamic management with vaginal delivery. I've seen the complications at 72 hours sometimes heralded with low platelets and by how much is hormonally driven and how much is hemodynamically driven.

### **Dr Zwicke:**

There can be spleen sequestration or the decreased counts known to occur with prostacyclin itself.

### **Dr Horn:**

Yes, all of the above. So that I actually like patients to remain in the ICU for the 72 hours after delivery.

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**Dr Zwicke:**

Always. We keep them there; 100% of people are in the ICU and it requires a re-education of the ICU nursing staff. They look well and act well. They're getting to the bathroom and they're walking around and they look fine. It's the potential for disaster; that's the reason they are there.

**Dr Horn:**

And then they can still crash.

**Dr Zwicke:**

And their typical crash is 3 am on morning 3. That's when the majority of deaths are reported. It's due to the massive fluid shifts back into the vascular tree. They develop acute pulmonary edema and are very difficult to rescue. This accounts for the majority of the pregnancy-related deaths in PAH patients.

**Dr Horn:**

Three days, absolutely. And some tell-tale signs; look at the platelets and LFTs.

**Dr Zwicke:**

Good point.

**Dr Levine:**

Absolutely, and I did want to get to that point. In the literature—especially in the older literature—there is a lot of controversy about the type of monitoring people should use. What

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are your thoughts regarding using PA catheter versus a CVP monitor during the delivery and postpartum in the ICU?

**Dr Oudiz:**

It's difficult to know whether you're treating the physician or you're treating the patient with invasive monitoring. Given the nature of all the issues we've just talked about, and the fact that things can creep up on you without being prepared, we tend to empirically place a PA catheter prior to delivery. In our center, our system works better with the surgical approach rather than a delivery in the CCU, which can be very impractical. So prior to any anesthesia or even an anesthesiologist's seeing the patient, we'll have the catheter already in the ICU and we'll keep it there until that critical period is over where we know that they have passed their highest risk of complications, somewhere around 72 hours. This brings up a question about the Pitocin and the induction of labor. Because we don't know all that well what happens relative to pulmonary vascular changes and pregnancy hormones in the peripartum period, and I think it's more than just fluid shifts that suddenly cause the right ventricle to acutely fail in the postpartum period.

**Dr Horn:**

Well, it's also the acute change – it's both the hemodynamic as well as hormonal –because we also have the acute change in the SVR.

**Dr Oudiz:**

Right. And so if there is a hormonal component to it, we don't know what it is, but it would intuitively involve for example, oxytocin, a vasoconstrictor, but also a hormone mediating the milk let-down response postpartum. So I worry a little bit about using Pitocin up front when it may be, at least in part, one of the reasons why patients do poorly postpartum.

**Dr Horn:**

I do something close to what Dianne does also. So we start the Pitocin, we do the epidural. We also aim for Monday, Tuesday and in our case the right heart cath goes in at the same

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time.

**Dr Zwicke:**

We used PA catheters initially, until we became more comfortable. Now, it's pretty much a CVP only. But, if there is any question, certainly put a PA catheter in. You can follow outputs for those patients that you are concerned with. I have found a limited right-heart echo each day more helpful as a diagnostic procedure.

**Dr Oudiz:**

Is the CVP monitoring enough for you to know that PVR is or isn't getting worse?

**Dr Zwicke:**

Yes. This is because I'm not treating the PVR, I'm treating the right ventricular function and I will have the echo tech come over after the epidural has worn off, about 4 hours after delivery, and do a right heart echo. I frequently use low-dose dobutamine to augment the RV systolic function during the periods of stress and volume shifts. The right ventricle has fast twitch muscle fibers that are extremely responsive to dobutamine. So low dose, 2- 4 mics, of dobutamine will augment your RV function tremendously. And especially in these post delivery. If you're concerned at all about how the RV looks—if it's a little sluggish to you— low-dose dobutamine goes a long way to help the diuresis and improve all the hemodynamics.

**Dr Horn:**

And when we do the CVP you can follow the right atrial saturation.

**Dr Zwicke:**

Right. One other thing to remember is that if the postpartum patient begins to desaturate and

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you haven't achieved the desired diuresis, they can blow open a previously closed PFO and begin to right to left shunt, causing systemic desaturation. This may also happen at the start of a dobutamine infusion, if it augments RV contractility, increasing the tricuspid regurgitation jet into the RA, opening the PFO. The treatment for both situations is aggressive IV diuresis. As soon as you decrease the intravascular volume, the shunting will decrease or stop.

**Dr Levine:**

Thanks, Dianne. Another area that we should discuss is counseling the patient regarding future pregnancies; that is, contraception, tubal ligation, etc..

**Dr Zwicke:**

Of the people we saw, we counseled 100% while we were following through the pregnancy. So, it was all set up for what type of contraception. We already knew if it was going to be an IUD, Depo, or a tubal ligation and when that was to occur. Necessary consents were already signed before they came in for induction. So, if an intended vaginal delivery converted to a C-section, it was all done. There were no last-minute decisions. It's all there on the chart. I think that the total preplanning is so important. I have cared for a lot of pregnant PAH patients; I'll be the first to advise against pregnancy, as the risk is still very high and we have a lot to learn yet. I do not tell patients that it's "okay" to get pregnant with pulmonary arterial hypertension. But, when they do become pregnant or are diagnosed during pregnancy, it is our job to properly and thoroughly counsel them, provide the best care we can, and provide all of the information we have available to us, so that they can make an informed decision in their situation.

**Dr Levine:**

Thank you. I think you've summed it up. I think that's the message that needs to hit home.

**Dr Zwicke:**

Well, it's a risk you don't need to take. All except one patient in our practice arrived pregnant and decided after counseling, to not abort. Since we are a referral center, these patients had

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already decided not to abort their pregnancies prior to arrival.. One intentionally, knowing she had pulmonary hypertension, treated at another center with PDE-5 inhibitors, came to our clinic in the 2nd trimester, unstable, and with mild right heart failure symptoms. She needed to convert to IV prostacyclin and delivered. She proceeded to become pregnant again after that despite all advice to not do so. She was treated for about a year with IV prostacyclins and converted to 2-drug oral therapy. She now has had a tubal ligation.

**Dr Oudiz:**

I would still consider her extremely lucky.

**Dr Zwicke:**

Yes, absolutely. So do I.

**Dr Horn:**

And so I think we would all agree on ending it exactly how Dianne ended it.

**Dr Levine:**

Agreed. Another point I believe we should touch on is where these patients are being managed. Everyone on the call today has had patients in this situation and has built multidisciplinary teams of specialists to help manage these patients when the need arises. These patients' care needs to be individualized and should be done at a specialty center with collaboration of the high risk OB, the anesthesiologist, the PAH specialist, and the rheumatologist (if needed)—everyone who is necessary to help coordinate the treatment of both the pregnancy and the PAH.

**Dr Zwicke:**



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I have just one other comment about the team. We leave out the respiratory therapy department frequently. If you are intending to proceed to a vaginal delivery, you can teach these moms to do a small breath Valsalva push without doing the usual deep and extended breath-hold type Valsalva. The normal delivery room nurse will instruct the patient, take a deep breath, bear down, hold until you turn blue. You can't do that with these patients. So, we actually set up training in our respiratory therapy department to teach the pregnant women how to push using their abdominal muscles and perineal muscles, instead of using the diaphragm. So, that would be one other thing to include in the pre-delivery teaching plan. It can be a partial forceps delivery, but they actually can do some pushing and they do shorter pushes using the abdominal muscles.

### **Dr Horn:**

I think that is something that is very important.

### **Dr Zwicke:**

But, we don't think about it until you're in the delivery room. The team sometimes needs to involve some of the people who have had more experience with some high forceps deliveries too.

### **Dr. Levine:**

I agree. Again, having an entire team involved before getting to this point is so important to be able to manage this or any scenario that may (and will) arise during pregnancy or delivery or in the post-partum period. There are, as we have seen today, individual centers that have acquired experience over time with these patients and have developed a team of experts working together when these women are referred. It is very important to have that experience behind us when taking care of these patients.

We are so fortunate to have had this whole group together today to discuss this complicated matter of PAH and pregnancy. This conversation only touches on how complicated and serious this situation can be and how many issues are involved. It also reflects on how little information (aside from case reports and case series) there are to help guide PAH specialists, obstetricians, and the whole team in managing these women when they are referred to us. This hour today has helped drive home the importance of bringing together each center's collective experience (albeit small) to help to develop guidelines as well as practical strategies to help better care for these women.

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I again would like to thank everyone for taking the time to join us today.

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