Transcript Details

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Shared Decision-Making in Pulmonary Arterial Hypertension: Strategies for PAH Treatment Initiation and Support

ReachMD Voiceover:
This is ReachMD, and the following Medical Industry Feature, titled, "Shared Decision-Making in Pulmonary Arterial Hypertension: Strategies for PAH Treatment Initiation and Support," is an educational activity brought to you by Johnson & Johnson. This program is intended for US healthcare professionals only and is not certified for continuing medical education. Please see full prescribing information for UPTRAVI® at www.UPTRAVIHCP.com, and the complete Important Safety Information at the end of this program.

UPTRAVI® (selexipag) is indicated for the treatment of pulmonary arterial hypertension (WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH. It's important to note that UPTRAVI® should not be taken by someone who takes gemfibrozil, because this medicine may affect how UPTRAVI® works and cause side effects, or if someone is allergic to selexipag or any of the other ingredients in UPTRAVI®.[1] Effectiveness of UPTRAVI® Tablets was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms.[1]

In consideration of the time spent participating in this activity, the speakers in this feature were paid an honorarium by Johnson & Johnson. The speakers are presenting on behalf of Johnson & Johnson. The patient featured in this video is a patient advocate through the SHARE Network, a program made up of people who are dedicated to inspiring others through their personal health journeys and stories of caring. She is a real UPTRAVI® patient with PAH and is sharing her personal experiences. Individual experiences may vary.

Now, here's your host, Lori Reed.

Lori:
Hi, I'm Lori Reed, and I'm an advanced practice provider. Today, we'll be discussing pulmonary arterial hypertension, also known as PAH, which is a rare form of pulmonary hypertension, or PH.[2] This progressive disease leads to right ventricular failure and death, which is why early diagnosis and treatment intervention are critical for helping improve patient outcomes.[2-3]

Also joining me today is Julie, who has been living with PAH since 2004. I'm looking forward to discussing her PAH journey and the role of shared decision-making.

Now, for some background, evidence-based guidelines, such as the 2022 European Society of Cardiology (ESC) and European Respiratory Society (ERS) Guidelines for the diagnosis and treatment of PH, provide the foundation for selecting proven therapies and help guide clinicians toward the most effective treatment approaches.[2]

In addition to the treatment guidelines, the PAH community came together in 2024 for the 7th World Symposium of Pulmonary Hypertension to review major advances in PAH management. This symposium produced a new expert consensus for the treatment of PAH for providers to also refer to.[4]

Prostacyclin pathway is one of three foundational PAH treatment pathways and targeting the prostacyclin pathway is an important strategy in effective disease management.[2]

The guidelines recommend adding UPTRAVI®, a prostacyclin receptor agonist (or PRA for short), at first follow-up for patients with idiopathic, heritable, or drug-induced PAH who present at intermediate–low risk of death while receiving endothelin receptor antagonists (ERA for short) and phosphodiesterase-5 inhibitors (PDE5i for short) therapy.[2] They also recommend UPTRAVI® for sequential dual-combination and triple-combination therapy with ERAs and/or PDE5is to reduce the risk of morbidity and mortality.[2]

The efficacy and safety of UPTRAVI® were evaluated in the phase 3 GRIPHON study, a multicenter, long-term, double-blind, placebo-controlled, parallel-group, event-driven study conducted in 1,156 patients with PAH. GRIPHON, the largest PAH pivotal trial, demonstrated that UPTRAVI® can delay disease progression and help reduce the risk of hospitalization for PAH.[1,5-6] The safety of UPTRAVI® was well established in the GRIPHON study. The adverse reactions occurring more frequently with UPTRAVI® compared with placebo by ≥3% were headache, diarrhea, nausea, vomiting, jaw pain, pain in extremities, myalgia, flushing, arthralgia, rash, anemia, and decreased appetite.[1] Adverse reactions were less frequent during the maintenance phase.[1] Please see the complete Important Safety Information continued at the end of this program.

Despite the availability of these evidence-based guidelines to help guide PAH care decisions, clinicians and patients may continue to face challenges when finding a treatment plan that fits within their lifestyle.[2,7] This is where shared decision-making becomes valuable, as it can help patients feel informed and educated about their treatment decision, to ensure that the choosen approach addresses both their specific concerns and their individual health needs.[8]

Shared decision-making in PAH care involves the collaborative dialogue where patients and healthcare providers discuss not only symptoms but also the broader impact of the disease and personal treatment goals. This respectful, two-way interaction helps establish common ground and supports informed, value-based decisions.[8]

Julie, in your experience, what are some challenges you observed early in your PAH journey when it came to starting treatment? And how would you describe your involvement in discussions and decisions about your treatment?

Julie:
I first began experiencing symptoms a few years before my diagnosis. I started feeling that it was hard to breathe, and I was then misdiagnosed with asthma, before an ER visit led me to testing and eventually my diagnosis.

Once I received my PAH diagnosis, we began looking at treatments. At the time, there weren't many available treatments, and I was concerned about being able to find a care plan that allowed me to keep up with my day-to-day activities.

This included my career as a real estate agent and being present for my three sons, niece and nephew.

I was really clear about my expectations for my therapy. I was able to be open and honest with my care team – which included a nurse practitioner like you, Lori – about my expectations and fears. Together, we were able to work to find something that worked for me.

Since my diagnosis, I've remained in close contact with my care team, who have helped me manage my PAH. They have supported me through every phase of my PAH journey.

Over time, this eventually led to a treatment regimen that included UPTRAVI®, so that I could be on an oral therapy targeting the prostacyclin pathway.[1]

Lori:
Thank you so much for sharing, Julie. Shared decision-making lays the foundation for patient engagement,[8] and it is important to recognize that disease management, particularly through a team-based approach, can play a critical role in determining therapeutic success.[9]

UPTRAVI® is titrated, usually weekly, to an individualized maintenance dose, also known as IMD, which means that every patient's dose may be different, and everyone does not need to reach the maximum dose to experience the potential benefit of UPTRAVI®.[1]

For example, recent data from the 10-year open-label extension (OLE) study of UPTRAVI® found that patients with a personalized low dose of UPTRAVI® (200 mcg to 400 mcg) achieved similar outcomes as patients with higher doses (up to 1600 mcg).[10] This long-term follow-up was conducted in patients who were treated with UPTRAVI® in the pivotal trial and OLE, which included 574 patients. Patients that were randomized to UPTRAVI® or the placebo in GRIPHON could enter the GRIPHON OLE either after experiencing a morbidity event during the double-blind treatment or at the end of the study.[11] Data from the GRIPHON study and its OLE reported survival among patients treated with UPTRAVI® for up to 10 years, including post hoc outcome assessments by individual maintenance dose, risk category, background therapy and time from diagnosis.[10] All analyses were descriptive only, baseline characteristics were not balanced.

These uncontrolled observations do not allow comparison with a control group not given UPTRAVI® and cannot be used to determine the long-term effect of UPTRAVI on mortality.[10]

It's important to note that side effects associated with UPTRAVI® use occur more frequently during the dose adjustment phase.[1] If a patient is experiencing side effects, their healthcare provider may tell them to change their dose, depending on what is right for them.[1]

Nurses working closely with individual patients and providing comprehensive supervision can also help PAH specialists start and maintain their patients on UPTRAVI®.[12] By extensive education on what to expect throughout the titration period, care teams can help patients achieve their IMD.[12]

Julie, can you tell me what it was like when you and your PAH care team first talked about starting UPTRAVI®? And what were your initial thoughts?

Julie:
I actually had my eye on the clinical trials of UPTRAVI®. When it became FDA-approved, I spoke to my care team about getting on it, and my care team helped me understand what to expect in those first few weeks. They also helped me manage some of my symptoms I was experiencing through the titration period.[1]

Lori:
We know that the titration period can be challenging for some people.[6] How did you and your PAH care team track how you were doing as you worked towards your maintenance dose? Did you create goals for yourself during this time?

Julie:
For tracking my progress towards my maintenance dose, I would go in every four months, and I usually do an echocardiogram about once a year.

During the titration period, my main goals were to stay on the therapy, feel comfortable with the process, and understand how increasing my dose might make me feel so I knew what to expect.

Lori:
Were there specific moments or support from your care team that helped you feel more confident staying on treatment?

Julie:
My care team during this period was great about keeping in touch; my nurse in particular. We keep an open line of communication to discuss how I am feeling, and she checks in with me as well.

You know, Lori, I'm curious. I've talked a lot about my relationship with my care team and how that impacted my journey. How do you approach open communication with your patients, to track how they're doing and help them set realistic goals?

Lori:
I prioritize building trust early on, so patients feel safe sharing how they're truly feeling—physically and emotionally. Open communication means listening actively and checking in regularly, not just during scheduled visits. I work with patients to co-create goals that reflect both clinical milestones and personal lifestyle needs. We use tools like symptom tracking, echocardiograms, and regular assessments to monitor patient progress and adjust goals as needed. It's important to me to meet patients where they are and help them feel empowered in their journey.

Now, as I mentioned before, data shows that adding UPTRAVI® to an appropriate patient's treatment regimen resulted in delayed disease progression and reduced the risk of hospitalization for PAH.[1,2,10]

The GRIPHON study is the largest PAH pivotal trial with the longest endpoint evaluation period of any prostacyclin pathway therapy.[5] The OLE of GRIPHON reported survival estimates of patients treated with UPTRAVI® for up to 10 years, including assessments by individual maintenance dose, risk category, background therapy and time from diagnosis, and long-term safety and tolerability.[10]

Indeed, long-term data from the pivotal study showed 5-year overall outcome of 74 percent with UPTRAVI®, which assessed 176 patients at risk at year 5, out of 574 patients at year 0.[10]

When looking at outcome rates based on a 4-strata risk category at UPTRAVI® initiation, the outcome rate at year 5 was more than 80 percent when UPTRAVI® was initiated in patients at low and intermediate-low risk. This included 188 patients at low risk and 266 patients at intermediate-low risk at the beginning of the study, with 63 and 85 patients respectfully still being evaluated at year 5.[10]

Notably, an 89 percent outcome rate at year 10 was observed when UPTRAVI® was initiated within six months of diagnosis as part of a triple combination therapy with an ERA plus a PDE5i. This included 23 patients at the beginning of the study, with 1 patient still being

evaluated at year 10. [10]

The safety profile of UPTRAVI® over this extended treatment period did not reveal new safety signals and was consistent with previous observations.[10]

Julie:
When I was first diagnosed with PAH in 2004, I remember doing my own research on the disease and learning about the short survival times.[13] Being here today to hear about decade long follow-up is remarkable. What are your thoughts on the amount of data available for UPTRAVI®?

Lori:
In my opinion, the depth and duration of data behind UPTRAVI® are reassuring—especially the 10-year survival data. Knowing UPTRAVI® treatment is backed by years of real-world and clinical experience also helps clinicians make informed decisions, especially when initiating therapy at first follow-up.

Julie:
Does this impact your decision-making when it comes to starting your patients on UPTRAVI®?

Lori:
Absolutely! The more we're informed, the better equipped we are to understand the potential impact of a therapy. In the case of UPTRAVI® as part of a proactive treatment strategy, it's a reminder that timely escalation, supported by strong evidence, has the potential to improve patient outcomes.

Julie, thank you for sharing your insights and experiences in your PAH journey. Your perspective highlights how early treatment escalation, strong patient/provider communication, and comprehensive support systems can impact PAH disease management.

As we wrap up, what would be your key advice or message for other PAH patients who might be starting their treatment journey or considering UPTRAVI® as part of their therapy?

Julie:
I want other people with PAH to have hope.

Since I reached my personal dose, I take one UPTRAVI® pill twice a day.[1] That's something that has really worked for me and my lifestyle. I don't feel tied down. I am enjoying my retirement. I can travel the country with my husband and live a pretty normal life.

I want to encourage other patients to work with their care teams, to look at all the options, and try something without fear.

Lori:
We hope this discussion reinforces the value of collaborative care approaches that empower patients to actively participate in their treatment decisions and achieve the best possible results.

Before we close, let's take a moment to review additional Important Safety Information.

ReachMD Announcer:

INDICATION

UPTRAVI® (selexipag) is indicated for the treatment of pulmonary arterial hypertension (PAH, WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH.

Effectiveness of UPTRAVI® Tablets was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms.

Patients had idiopathic and heritable PAH (58%), PAH associated with connective tissue disease (29%), and PAH associated with congenital heart disease with repaired shunts (10%).

IMPORTANT SAFETY INFORMATION

CONTRAINDICATIONS
Concomitant use of strong inhibitors of CYP2C8 (eg, gemfibrozil) with UPTRAVI® is contraindicated.

Hypersensitivity to the active substance or to any of the excipients is contraindicated.

WARNINGS AND PRECAUTIONS

Pulmonary Edema with Pulmonary Veno-Occlusive Disease (PVOD)

Should signs of pulmonary edema occur, consider the possibility of associated PVOD. If confirmed, discontinue UPTRAVI®.

ADVERSE REACTIONS

Adverse reactions more frequent compared to placebo (≥3%) seen with UPTRAVI® tablets are headache (65% vs 32%), diarrhea (42% vs 18%), jaw pain (26% vs 6%), nausea (33% vs 18%), myalgia (16% vs 6%), vomiting (18% vs 9%), pain in extremity (17% vs 8%), flushing (12% vs 5%), arthralgia (11% vs 8%), anemia (8% vs 5%), decreased appetite (6% vs 3%), and rash (11% vs 8%).

These adverse reactions are more frequent during the dose titration phase.

Hyperthyroidism was observed in 1% (n=8) of patients on UPTRAVI® tablets and in none of the patients on placebo.

DRUG INTERACTIONS

CYP2C8 Inhibitors

Concomitant administration with gemfibrozil, a strong inhibitor of CYP2C8, doubled exposure to selexipag and increased exposure to the active metabolite by approximately 11-fold. Concomitant use of UPTRAVI® with strong inhibitors of CYP2C8 is contraindicated.

Concomitant administration of UPTRAVI® with clopidogrel, a moderate inhibitor of CYP2C8, had no relevant effect on the exposure to selexipag and increased the exposure to the active metabolite by approximately 2.7-fold. Reduce the dosing of UPTRAVI® to once daily in patients on a moderate CYP2C8 inhibitor.

CYP2C8 Inducers

Concomitant administration with an inducer of CYP2C8 and UGT 1A3 and 2B7 enzymes (rifampin) halved exposure to the active metabolite. Increase UPTRAVI® dose, up to twice, when co-administered with rifampin. Reduce UPTRAVI® when rifampin is stopped.

DOSAGE AND ADMINISTRATION

Recommended Dosage

Recommended starting dose is 200 mcg twice daily for UPTRAVI® tablets. Tolerability may be improved when taken with food. Increase by 200 mcg twice daily, usually at weekly intervals, to the highest tolerated dose up to 1600 mcg twice daily. If dose is not tolerated, reduce to the previous tolerated dose.

Patients With Hepatic Impairment

For patients with moderate hepatic impairment (Child-Pugh class B), the starting dose of UPTRAVI® tablets is 200 mcg once daily. Increase by 200 mcg once daily at weekly intervals, as tolerated. Avoid use of UPTRAVI® in patients with severe hepatic impairment (Child-Pugh class C).

Co-administration With Moderate CYP2C8 Inhibitors

When co-administered with moderate CYP2C8 inhibitors (eg, clopidogrel, deferasirox and teriflunomide), reduce the dosing of UPTRAVI® to once daily.

Dosage Strengths

UPTRAVI® tablet strengths:
200, 400, 600, 800, 1000, 1200, 1400, and 1600 mcg.

Additional Important Safety Information for UPTRAVI® IV

Use UPTRAVI® for injection in patients who are temporarily unable to take oral therapy.

Administer UPTRAVI® for injection twice daily by intravenous infusion at a dose that corresponds to the patient's current dose of UPTRAVI® tablets (see Table 1 in full Prescribing Information). Administer UPTRAVI® for injection as an 80-minute intravenous infusion.

Adverse Reactions: Infusion-site reactions (infusion-site erythema/redness, pain and swelling) were reported with UPTRAVI® for injection.

Please see full Prescribing Information at UPTRAVIHCP.com.

cp-126160v8

ReachMD Announcer:
This program was sponsored by Johnson & Johnson. If you missed any part of this discussion, visit Industry Features on ReachMD.com, where you can Be Part of the Knowledge.

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