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"//connect.facebook.net/en_US/all.js#xfbml=1"; fjs.parentNode.insertBefore(js, fjs);
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Section Editor: **Martha Kingman, NP**

Women with idiopathic pulmonary arterial hypertension (IPAH) are often of childbearing age and outnumber males with IPAH by a 4:1 ratio.¹ All patients with PAH should have a primary care provider, but in some cases the PH provider may be the only health care professional that the PH patient is seeing on a regular basis. Therefore, while the primary focus is on the treatment of PH, routine preventive care should also be incorporated. This article will examine important preventive health care strategies for patients with PH, with a special focus on women's issues. Topics covered include immunization; counseling on tobacco, alcohol, and drug avoidance; screening for osteoporosis, breast cancer, cervical cancer, sexually transmitted disease, and colorectal cancer; and pregnancy prevention. A summary of select U.S. Preventive Services Task Force (USPSTF) screening recommendations is listed in Table 1.

Table 1:

Summary of UPSTF Recommendations That Commonly Apply to PH Patients

Screening for breast cancer	Women age 50 and over
Screening for cervical cancer with Pap smear	Women age 21 to 65 who are sexually active and have a cervix
Screening for Chlamydia	Women age ≥ 25 or at increased risk
Screening for colorectal cancer	Adults age 50 to 75; generally discontinue when life expectancy <10 years and avoid screening when life expectancy <5 years.
Screening for osteoporosis	Adults age >65 or at increased risk.
Screening for tobacco, alcohol, and drug use	All adults: even brief counseling increases quit rates for tobacco and reduces alcohol consumption
Depression	All adults: screening plus referral for therapy reduces the burden of suffering from depression
Immunizations	Influenza, varicella or zoster, tetanus/diphtheria and pneumovax are recommended

Standard screening tests, performed at specified intervals, can be a cost-effective way to identify and treat potential health problems before they develop or worsen.² However, while many of the usually recommended assessments apply, the presence of PH leads to modifications in some cases. For example, pneumococcal vaccination is recommended at age 65 or with “chronic disease,” and thus all patients with PH should receive the pneumococcal vaccine. Additionally, if the first pneumococcal vaccination is received before the age of 65, then a second dose should be given at or after age 65. Other vaccination recommendations include annual influenza vaccination and a one-time zoster vaccination for patients 60 years and over.

Similarly, colorectal screening should probably only be considered in patients with very stable PAH, as current recommendations are for screening only in patients age 50 to 75 with a life expectancy greater than 10 years.³ Screening options include fecal occult blood testing, sigmoidoscopy, and colonoscopy, all of which have demonstrated a mortality benefit in other patient populations.³ The type of screening should also be carefully considered: colonoscopy is typically a preferred test in the general population, but it requires significant sedation which can pose a risk for PH patients. In addition to the other screening options above, CT-colonography is now also a recommended option that requires bowel preparation but no sedation.³ Therefore, while the lack of sedation makes this option attractive, the tolerability of the large amount of oral fluid needed for bowel preparation needs to be considered.

Preventive care recommendations that should be given to all patients with PAH include counseling on tobacco, alcohol, and drug use avoidance.⁴ Patients should also be screened for depression, and if present, referred for treatment. Several depression screening tools are available and easy to use; or alternatively, asking two simple questions may be as effective as longer questionnaires: “Over the past two weeks, have you felt down, depressed, or hopeless?” and “Over the past two weeks, have you felt little interest or pleasure in doing things?”⁵

Women with PH also need routine well-woman screening. After menopause, women are at increased risk of developing osteoporosis.⁶ Some women with PAH associated with connective tissue disease may be taking prednisone, which is also a risk factor for the development of osteoporosis. Bone mineral density measurement using dual-energy x-ray absorptiometry (DXA) of the hip and lumbar spine and quantitative ultrasonography of the calcaneus (a noninvasive test) can help identify early manifestations of the disease. This test is recommended for anyone age 65 or older and those at risk for developing osteoporosis.⁶ A healthy diet with adequate amounts of calcium and vitamin D can help prevent osteoporosis. Experts recommend premenopausal women consume at least 1000 mg of calcium and 400 to 600 international units of vitamin D per day. Postmenopausal women should have an intake of 1200 mg of calcium and 800 international units of vitamin D daily.⁷

The American College of Physicians (ACP) and the USPSTF recommend screening mammography be conducted biennially in women ages 50 to 74 years, and recommend mammography be considered in women ages 40-49 years, after discussing the risks and benefits.⁸ The benefit of screening mammography in patients under age 50 is a potential decrease in breast cancer mortality of approximately 15%, but at the cost of potentially false-positive results, need for biopsies, and radiation exposure.⁹

The lack of evidence to support *annual* cervical screening has led to changes in testing frequency. The American Congress of Obstetricians and Gynecologists (ACOG) recommend women have their first cervical cancer screening at age 21 and once every 2 years until age 30, though women at increased risk due to immunosuppression, prior cervical cancer, or prior abnormal cytology results may need more frequent screening.¹⁰ Women older than 30 years of age with 3 consecutive negative cervical cytology results may have screening with either Pap test or liquid-based cytology every 3 years.¹⁰ Routine cervical screening should be discontinued in women who have had a total hysterectomy for noncancerous causes regardless of age, and discontinuation can also be considered in women over age 65 who have had 3 consecutive negative cytology results and no abnormal results in the last 10 years.¹⁰ These guidelines also apply to women who have been vaccinated against the human papillomavirus.¹⁰ Women ages 25 years or younger and all women who have new or multiple sex partners should also be screened for Chlamydia trachomatis, a common sexually transmitted disease that can lead to pelvic inflammatory disease.¹¹ This screening process also provides an opportunity to counsel patients on safe sex practices.

The physiological, cardiovascular, and pulmonary changes that occur during pregnancy have been associated with high maternal mortality in PAH, and thus pregnancy avoidance and use of adequate contraception are important.^{12,13} Additionally, one of the most common categories of medications to treat PAH, the endothelin receptor antagonists (ERAs), are category X in pregnancy, as they caused severe birth defects in animal studies.¹⁴ Pregnancy must be excluded prior to initiating therapy and women must be educated about the risks, required monthly pregnancy testing, and need for adequate contraception (Table 2; see also Pregnancy article and Contraception in PH article for additional details).^{15,16} Women should be advised to notify their health care professional of any delay in onset of menses or any other reason to suspect pregnancy, so that immediate pregnancy testing can be performed and the ERA discontinued.^{15,16}

Table 2:

Acceptable Methods of Contraception for Patients on ERAs

Methods to Use Alone	Combination Methods	
	Hormone Methods Choose one and use with a barrier method	Barrier Methods Use both OR choose one and use with a hormone method
Intrauterine devices (IUDs)	Estrogen and progesterone	
<ul style="list-style-type: none"> • Copper T 380A IUD • LNg 20 IUS (progesterone IUD) • Tubal sterilization 	<ul style="list-style-type: none"> • Oral contraceptives • Transdermal patch • Vaginal ring • Progesterone only • Injection • Implant 	<ul style="list-style-type: none"> • Diaphragm with spermicide OR • Cervical cap with spermicide • Male condom (with or without spermicide)

A partner's vasectomy still requires 1 additional method of contraception

Actelion. Important Safety Information about Tracleer. <http://www.tracleer.com/About-Tracleer-Safety-Information>. Accessed August 31, 2011.

Gilead. Letairis Product Information. http://www.gilead.com/pdf/letairis_pi.pdf. Accessed August 31, 2011.

Finally, although we began this article with the suggestion that the PH provider might consider tackling some or all of their patient's preventive health care needs, in reality, the large number of preventive care items required is a strong argument for encouraging all patients to establish good primary care as well. Studies have shown that patients with a primary care physician are more likely to receive appropriate preventive care and may have improved survival.¹⁷ As patients with PAH are living longer, the need for routine health screening becomes more important and should be incorporated into the plan of care in order to identify other potential health problems. The variation in recommendations supports the need for individualized discussions with and involvement of the patient in understanding the risks and benefits of certain screening tests.

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Javier Jimenez, MD

PH Clinicians & Researchers Member
Director of Advanced Heart Failure and Pulmonary Hypertension
South Miami Hospital
Miami, Florida

