Pulmonary Hypertension & Congenital Heart Disease (CHD)
PULMONARY HYPERTENSION, or PH, is complex and often misunderstood. PH means high blood pressure in the lungs and is different from regular hypertension. In PH, the blood vessels in the lungs become damaged and/or narrowed and the heart has to work harder to pump blood through them.

PH can be caused by many different problems such as heart failure, diseases that damage the lungs or multiple hardened blood clots. It is important to understand that not all PH is the same.

PH affects people of all ages and ethnic backgrounds. The most common symptoms are shortness of breath with physical activity, fatigue, lightheadedness and sometimes fainting. Because these symptoms can be caused by any number of other medical problems, diagnosis is often delayed. Identifying a case of PH can be difficult and may require a specialist. Once the type of PH is diagnosed, treatment can begin immediately.

One form of PH is called pulmonary arterial hypertension (PAH). PAH is a complex, progressive type of PH where the high blood pressure in the lungs occurs because tiny blood vessels that carry blood through the lungs (pulmonary arteries) are narrowed, thickened and stiff. As PAH advances, the heart may lose its ability to pump enough blood through the lungs to meet the needs of the body.

There are several types of PAH. Idiopathic PAH (IPAH) is PAH without any other obvious medical problem leading to high blood pressures in the lung. Heritable PAH (HPAH) comes from abnormal genes that cause PAH. Heritable PAH may be passed on to some members of your family. PAH can be associated with other medical conditions such as connective tissue diseases (scleroderma and lupus...
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PAH is a serious condition, and without treatment, symptoms will become worse, leading to heart failure and even death. Proper diagnosis and treatment from a doctor who understands PAH is essential. Every patient is different. The choice of treatment is based on how sick a patient is and the risks and benefits of any particular therapy. Regardless of risk, it is important that patients and their health care providers engage in frequent follow ups with ongoing discussions about the management of their condition. Current guidelines suggest that changes in therapy should be considered for patients not reaching their treatment goals.

While no cure has yet been found for PAH, increased research has resulted in treatments that allow patients to live longer, fuller lives with far less interference from the disease. Even more promising research is being conducted every day that is not only advancing our understanding of the PAH disease, but also potentially identifying new treatment options for patients in the future.
What is Congenital Heart Disease?

Congenital heart disease (CHD) can be a number of heart and blood vessel abnormalities that develop during fetal development prior to a person's birth. These abnormalities are often split into two categories: “simple” (a single abnormality of the heart valves, chambers or major vessels) or “complex” (multiple abnormalities of the heart valves, chamber or major vessels).

CHD is not a common condition. It affects less than one percent of the population. Today, people with CHD are living much longer lives because of recent improvements in surgery and treatment. In fact, there are now more adults — more than one million in the US — than children with CHD.

How is CHD related to PH?

Many types of CHD can lead to pulmonary hypertension (PH). About 10 percent of people with CHD will develop PH. The development of PH in people with CHD may worsen their symptoms and shorten their life, so it’s important to get properly diagnosed and treated for both PH and CHD.

PH usually develops in people with CHD because of increased blood flow into the lung vessels. This flow often comes from defects or “shunts” in the heart or the major vessels.

Common examples include holes between the upper heart chambers (atrial septal defects) or the lower heart chambers (ventricular septal defects). In these cases, blood flows from the left heart into the right heart and then into the lung vessels. If there is enough extra flow for a long enough period of time, the lung vessels start to become thicker and stiffer, and the pressure within them begins to increase.

If the pressure in the lung vessels increases enough, the flow across the defect in the heart will start to move in the opposite direction. This results in blood with low oxygen entering the body’s arterial circulation, lowering the overall oxygen level and causing cyanosis (blue fingers and lips). This is known as Eisenmenger syndrome. It’s important to detect heart defects and shunts as early in life as possible, so that they can be corrected and PH can be avoided.

Fortunately, recent evidence suggests that current PH treatments, coupled with other therapies, can help people who have developed PH-related CHD.

How is PH detected in people with CHD?

A cardiologist or a CHD specialist should be able to detect the development of PH. If someone with CHD notices an increase or worsening of symptoms (shortness of breath, intolerance to exercise, poor energy or bouts of lightheadedness or fainting), this will alert the physician to the possibility of PH. A physical exam consisting of an echocardiogram or cardiac ultrasound is an important test to see if PH may be developing.

A cardiac catheterization is the most definitive test for PH. This allows your doctor to directly measure pressure in the lung vessels. Catheterization also allows for a doctor to determine the type of PH the patient has, which will help them recommend the best therapy.

What can be done if someone develops CHD-associated PH?

People with CHD-associated PH should be watched closely and regularly by doctors in centers with experience managing the condition. This is especially true for people with Eisenmenger syndrome.
In general, people with CHD-associated PH benefit from a team-based approach to care with input from both a CHD specialist and a PH specialist. There are also several specific treatment options that should be considered:

**Oxygen supplementation:** Many people with CHD-associated PH have low oxygen levels in the blood. Some also have different degrees of lung disease. While oxygen therapy has not been definitively shown to improve exercise tolerance or survival, it may make symptoms better.

**Exercise:** Most people with CHD-associated PH benefit from regular, low-to-moderate intensity exercise, such as walking. Care should be taken to avoid high intensity exercise as it is usually not well tolerated by patients.

**Anticoagulation:** Anticoagulants or blood thinners likely provide little benefit to most people with CHD-associated PH. Many people with CHD-associated PH are at increased risk of bleeding, so the level of anticoagulation also must be watched very carefully.

**Diuretics:** People with CHD-associated PH can develop fluid build up in their abdomen or legs. When this happens, diuretics can be used to help control the access of fluid. Care must be taken to avoid overuse of these medicines, because it can cause dehydration, which in turn can cause low blood pressure and a dangerous thickening of the blood.

**Treatment of iron deficiency:** The low blood oxygen levels that people with Eisenmenger syndrome may experience can cause the body to increase its blood count. Blood draws (phlebotomy) may or may not be recommended to avoid excessive thickness of the blood. In either case, iron deficiency is fairly common and should be addressed to avoid anemia.

**Avoidance of pregnancy:** CHD-associated PH carries an increased risk for both mother and child during pregnancy. With Eisenmenger syndrome, there is a 50 percent risk of maternal death and a 40 percent risk of miscarriage. Women with CHD-associated PH should avoid pregnancy and discuss birth control options with their doctors.

**Advanced medical therapy:** People with CHD-associated PH should be considered for PH medicines. People with this condition have even participated in a number of clinical studies of these medicines.

The BREATH-5 study specifically showed the benefit of bosentan (an endothelin antagonist) in people with Eisenmenger syndrome. The two other major classes of PH medicines (phosphodiesterase-5 inhibitors and prostanoids) have also shown benefit in people with CHD-associated PH. Such medicines should be used under the supervision of a PH specialist.

**Transplant:** If people with CHD-associated PH do poorly despite medical therapy, they may be considered for lung or combined heart and lung transplantations. Those undergoing isolated lung transplantation may need to have surgical repair of their CHD at the same time, if not previously repaired. People with CHD-associated PH should be referred early for consideration of transplantation to allow time for a complete evaluation.
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¿Qué es la enfermedad cardíaca congénita?
La enfermedad cardíaca congénita (ECC) está constituida por un número de anormalidades del corazón y los vasos sanguíneos que aparecen durante el desarrollo fetal antes del nacimiento de una persona. Estas anomalías se dividen frecuentemente en dos categorías: “simple” (una sola anormalidad de las válvulas del corazón, cámaras o grandes vasos) o “complejas” (múltiples anormalidades de las válvulas del corazón, cámaras o grandes vasos).

La ECC no es una condición común. Afecta a menos del uno por ciento de la población. Hoy, las personas con ECC están teniendo una vida mucho más larga debido a las mejoras recientes en la cirugía y el tratamiento. De hecho, ahora hay más adultos —más de un millón en los Estados Unidos— que niños con ECC.

¿Cómo se relaciona la ECC con la hipertensión pulmonar?
Muchos tipos de ECC pueden conducir a hipertensión pulmonar (HP). Cerca de 10 por ciento de las personas con ECC desarrollarán HP. El desarrollo de HP en las personas con ECC puede empeorar sus síntomas y acortar sus vidas, por lo que es importante ser diagnosticados y tratados correctamente tanto para la HP como para la ECC.

La HP se desarrolla generalmente en personas con ECC debido al aumento del flujo sanguíneo hacia los vasos pulmonares. Este flujo viene a menudo de defectos o “cortocircuitos” en el corazón o los grandes vasos.

Ejemplos comunes incluyen orificios entre las cámaras cardíacas superiores (defectos del septo interauricular) o las cámaras cardíacas inferiores (defectos del septo ventricular). En estos casos, la sangre fluye desde el corazón izquierdo hacia el corazón derecho y luego hacia los vasos pulmonares. Si hay suficiente flujo extra durante un período de tiempo suficientemente largo, los vasos pulmonares empiezan a volverse más gruesos y rígidos, y la presión dentro de ellos comienza a aumentar.

Si la presión en los vasos pulmonares aumenta lo suficiente, el flujo a través del defecto en el corazón comenzará a moverse en la dirección opuesta. Esto determina que la sangre con oxígeno bajo entre en la circulación arterial del cuerpo, disminuyendo el nivel total del oxígeno y causando cianosis (dedos y labios azules). Esto se conoce como síndrome de Eisenmenger. Es importante detectar los defectos cardíacos y los cortocircuitos tan temprano en la vida como sea posible, para que puedan ser corregidos y se pueda evitar la HP.

Afortunadamente, la evidencia reciente sugiere que los tratamientos actuales para la HP, junto con otras terapias, pueden ayudar a las personas que han desarrollado la ECC relacionada con HP.

¿Cómo se detecta la HP en personas con ECC?
Un cardiólogo o un especialista en ECC debe ser capaz de detectar el desarrollo de HP. Si alguien con ECC observa un aumento o empeoramiento de los síntomas (dificultad para respirar, intolerancia al ejercicio, falta de energía o ataques de mareos o desmayos), esto alertará al médico sobre la posibilidad de HP. Un examen físico debe incluir un ecocardiograma o ecografía cardíaca que son pruebas importantes para ver si la HP puede estar desarrollándose. Un cateterismo cardíaco es la prueba más definitiva para la HP. Esto permite que su médico mida directamente la presión en los vasos pulmonares. El cateterismo también permite que un médico determine el tipo de HP que tiene el paciente, lo que les ayudará a recomendar la mejor terapia.

¿Qué puede hacerse si alguien desarrolla una ECC asociada con HP?
Las personas con ECC asociada con PH deben ser observadas de cerca y regularmente por los médicos en los centros con experiencia en el manejo de la condición. Esto es especialmente cierto para las personas con síndrome de Eisenmenger.
En general, las personas con ECC asociada con HP se benefician de un enfoque basado en el equipo de atención con la contribución de un especialista en ECC y un especialista en HP. También hay varias opciones de tratamiento específico que deben considerarse:

**Administración de oxígeno:** Muchas personas con ECC asociada con HP tienen bajos niveles de oxígeno en la sangre. Algunos también tienen diferentes grados de enfermedad pulmonar. Aunque no se ha demostrado definitivamente que la oxigenoterapia mejore la tolerancia al ejercicio o la supervivencia, puede mejorar los síntomas.

**Ejercicio:** La mayoría de las personas con ECC asociada con HP se benefician de ejercicios regulares de intensidad baja a moderada, como caminar. Se debe tener cuidado de evitar el ejercicio de alta intensidad, ya que por lo general no es bien tolerado por los pacientes.

**Anticoagulación:** Los anticoagulantes o diluyentes sanguíneos probablemente proporcionen pocos beneficios a la mayoría de las personas con ECC asociada con HP. Muchas personas con ECC asociada con HP tienen un mayor riesgo de sangrado, por lo que el nivel de anticoagulación también debe ser observado con mucho cuidado.

**Diuréticos:** Las personas con ECC asociada con HP pueden desarrollar acumulación de líquido en el abdomen o en las piernas. Cuando esto sucede, se pueden utilizar los diuréticos para ayudar a controlar el exceso de líquidos. Se debe tener cuidado de evitar el uso excesivo de estos medicamentos, ya que pueden causar deshidratación, que a su vez puede causar presión arterial baja y un engrosamiento peligroso de la sangre.

**Tratamiento de la deficiencia de hierro:** Los bajos niveles de oxígeno en la sangre que las personas con síndrome de Eisenmenger pueden experimentar pueden hacer que aumente el recuento sanguíneo corporal. Los drenajes sanguíneos (flebotomía) pueden o no recomendarse para evitar un engrosamiento excesivo de la sangre. En cualquier caso, la deficiencia de hierro es bastante común y debe ser abordada para evitar la anemia.

**Prevención del embarazo:** La ECC asociada con HP conlleva un mayor riesgo tanto para la madre como para el niño durante el embarazo. Con el síndrome de Eisenmenger, existe un riesgo del 50 por ciento de muerte materna y un riesgo del 40 por ciento de aborto espontáneo. Las mujeres con ECC asociada con HP deben evitar el embarazo y discutir opciones de control de la natalidad con sus médicos.

**Tratamiento médico avanzado:** Las personas con ECC asociada con HP deben ser consideradas para los medicamentos de HP. Las personas con esta condición han incluso participado en una serie de estudios clínicos de estos medicamentos.

El estudio BREATH-5 mostró específicamente el beneficio de bosentan (un antagonista de la endotelina) en las personas con síndrome de Eisenmenger. Las otras dos clases principales de medicamentos para la HP (inhibidores de la fosfodiesterasa-5 y prostanoides) también han mostrado beneficios en las personas con ECC asociada con HP. Estos medicamentos deben utilizarse bajo la supervisión de un especialista en HP.

**Trasplante:** Si las personas con ECC asociada con HP no evolucionan bien a pesar del tratamiento médico, pueden ser consideradas para trasplantes pulmonares o trasplantes combinados de corazón y pulmón. Aquellos que se someten a un trasplante de pulmón aislado pueden necesitar una reparación quirúrgica de su ECC al mismo tiempo, si no se reparó previamente. Las personas con enfermedad ECC asociada a HP deben ser remitidas temprano para considerar el trasplante para dar tiempo a una evaluación completa.
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