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**Social Security Administration
Compassionate Allowances Outreach Hearing on
Cardiovascular Disease and Multiple Organ Transplants
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Thank you for the opportunity to present the views of the American Heart Association and its division, the American Stroke Association. I am Dr. Ileana Piña, Professor of Medicine at Case Western Reserve University. I am also a member of the Scientific Advisory and Coordinating Committee of the American Heart Association and a member of the Councils on Clinical Cardiology and Quality.

AHA is the nation's largest voluntary health organization, with over 22 million volunteers and supporters. Since 1924, AHA has dedicated itself to building healthier lives free of cardiovascular disease and stroke – the #1 and #3 leading causes of death in the United States – through research, public and provider education, healthcare provider quality improvement programs, and advocacy.

AHA appreciates the Social Security Administration's decision to examine the compassionate allowance conditions and to consider what forms of cardiovascular disease should be added to the list. Unfortunately, CVD is a very common disease; an estimated 81 million American adults – more than one in three – have one or more types of cardiovascular disease. CVD, which includes high blood pressure, coronary heart disease, heart failure, stroke, and congenital cardiovascular defects, can vary in type and severity. CVD can also vary in its impact on the patient. For some patients with severe forms of CVD, cardiovascular disease can make it difficult, if not impossible, to lead a normal life. CVD is also an equal opportunity disease since it is also the number one cause of death in women,

As a physician, I've seen patients who are unable to work, afford compliance with recommended medical care, or even engage in the simplest activities of daily living because of CVD. And I've also seen some of those same patients denied disability benefits or have to apply more than once before being approved. And as you know, once approved, they still have to wait 24 months before becoming eligible for Medicare. By the time eligibility occurs, some of these patients will have already had many events, such as hospitalizations and some will have died. For those patients, the length of the application process and Medicare's waiting period are deadly. So I encourage the Administration to consider adding certain types of CVD to the compassionate allowances list. Revisions to the overall disability process may be warranted based on the patient examples I'll provide today.

Heart Failure

The first form of CVD I'll discuss is heart failure, often called congestive heart failure, although congestion only touches the tip of the iceberg.

Heart Failure is often the final pathway of a multitude of heart disorders.

Heart failure, a chronic and progressive condition seen in childhood as well as adulthood, is probably one of the worst sequelae of any disease of the heart, whether of the heart valves, the coronary arteries, or a problem of the heart muscle itself. It is a condition marked by many hospitalizations and high mortality.

5.8 million Americans have heart failure.

It is important to understand that, while CVD includes a wide spectrum of conditions and treatment options, can be the secondary result of other conditions and diseases, and can be treated, with an improvement in symptoms; at its most severe, the heart failure that often results **cannot be cured**. At best, the symptoms are managed with some maintenance of quality of life. This is the point of disability where compassionate allowances should be a part of the options available for these patients.

Across the U.S., heart failure is marked by a significant number of hospital readmissions. It is the most common cause of readmission within 30 days of discharge, and the mean length of stay for heart failure is 4.5 days. It has been estimated that approximately 2/3 of hospital readmissions are preventable. One of five patients are likely to be rehospitalized within 30 days and by 6 months, 50% have been rehospitalized.

Heart failure readmissions change the patients' mortality factors so that at 12 months, their mortality risk is about 30%, simply because they have been hospitalized. Half of these patients are dead within 5 years. This gives heart failure a prognosis worse than every major cancer, with the exception of pancreatic and lung cancer. Certainly there is a tremendous opportunity to improve upon this natural history, and improve outcomes for patients hospitalized with heart failure.

Symptoms of heart failure can include shortness of breath, swelling in the feet, ankles, legs or abdomen or weight gain, tiredness, fatigue, persistent coughing or wheezing, lack of appetite, nausea, confusion, impaired thinking, or increased heart rate. At the most severe level, these symptoms are present not just with exertion, but even at rest.

Patients with advanced heart failure, New York Heart Association Class III (marked limitations even during less-than-ordinary activity; comfortable only at rest) and Class IV (severe limitations; experience symptoms even while at rest), and patients with chronic angina that occurs at low workloads, typically cannot work due to the severity of the symptoms and the medical care they need.

Although the NYHA Classification is useful clinically, it is subjective based on both the clinician and the patient's perspective of activity function. We do use more objective means of

measurement for both the heart's function and the patient's activity and functional capacity (their ability to engage in activity).

Testing for LV Function

Cardiac testing is available to indicate LV function and can be determined by echo and by other imaging techniques, e.g., MRI.

The imaging modality techniques are complementary when diagnosing cardiovascular disease

- Transthoracic Echo for assessing ventricular function and valvular regurgitation
- Doppler techniques for stenosis of valves
- MRI spares radiation but cannot be used if pacemakers/ICDs are in place
- Only invasive testing with cardiac catheterization can obtain true hemodynamics

Measuring B-type Natriuretic Peptide (BNP) and N-terminal Pro BNP (NT-Pro-BNP) have proven to be predictive of mortality, cardiovascular events, AND overall prognosis in heart failure. However they are highly variable between patients making standardization difficult. They lack specificity, and false positive elevations do occur. More research is needed in this area.

Functional Capacity Determination

Factors that predict mortality, such as pump function, do not correlate with actual patient ability to perform activity, i.e., function or functional capacity, the ability to do activity. Yet functional capacity is a powerful predictor of mortality.

There are a variety of clinical conditions that lead to end stage heart failure, yet in the late stages of this disease, evaluation of the patient's condition and prognosis is similar. Determining the function of the total patient requires taking a good history and testing by exercise with or without cardiopulmonary gas measurement. The most accurate assessment is done with cardiopulmonary exercise testing. The 6 minute walk can be used to determine function of sicker patients at a low level comparable to daily living activities. Some advanced heart failure patients are too ill to perform these examinations due to dependence on inotropes or need for short term mechanical circulatory support. For patients with end-stage heart failure, some will qualify for advanced heart failure surgical therapies, but many more will be referred for palliative care. Accordingly heart failure patients, who despite attempts at medical therapy, have 6-min walk distance less than 309 feet, VO2 max less than 50% of predicted, or are dependent upon inotrope therapy or mechanical support have a extremely high risk of mortality and should be considered for Compassionate Allowance.

Prognosis for Improvement

We know that exercise (as provided and taught in cardiac rehabilitation programs) can improve quality of life for many heart failure patients and can have a modest beneficial effect on outcomes. Yet cardiac rehabilitation for these patients is not always available nor is it covered by Medicare or by the majority of insurers.

There are no reliable scales or measures that assess the prognosis for improvement for heart failure. As previously noted, the NYHA classification level is very subjective. There are health

status scales which include quality of life that appear to capture the adequacy (or lack thereof) of capacity for daily living. However, few clinicians use any of these questionnaires (instruments) in their practice. We need better measures that are clinically accessible and truly prognostic. Hence the testing of the heart and the patient are what we rely on today to assess their future outcome.

Congenital Cardiovascular Heart Disease

The next form of CVD I'd like to discuss is congenital cardiovascular disease. In nine out of every 1,000 births, some form of congenital heart disorder will be present. Defects range in severity from simple problems, such as "holes" between chambers of the heart, to very severe malformations, such as complete absence of one or more chambers (hypoplastic left heart syndrome, single ventricle) or valves (aortic, tricuspid or pulmonary atresia). Congenital heart disease can also increase the risk of developing certain other medical conditions, including pulmonary hypertension, arrhythmias, infective endocarditis, heart failure, and the need for many to take blood thinners (anticoagulant) medications.

For more complex lesions, limitations are common. These children have severe functional limitations, require frequent office visits, suffer frequent hospitalization and have very complicated medicine regimens. Often full time caregiver support is needed. Psychosocial support is critical. Some children with congenital heart disease have developmental delay or other learning difficulties and can only attend school part-time, and some progress to require heart transplantation.

As with adults, all of the same tools to evaluate cardiac function are available for adolescents and older children, however full exercise stress testing with cardiopulmonary function testing cannot be applied to children less than eight years of age and older children with small body builds; thus we don't have the usual exercise endurance time and peak oxygen consumption data available for young children and infants to use as a prognostic indicator. In addition, nuclear testing is not readily available at most children's hospitals, thus patients with concerns about ischemic heart (coronary) disease, such as those with Kawasaki disease or a history of coronary surgery as part of their repair (e.g. transposition), or other lesions where potential coronary artery injury is a possibility may have more difficulty having dobutamine/exercise or nuclear stress testing performed for follow-up and evaluation.

The symptomatic progression of adults with congenital heart disease can be quite variable. Patients with single ventricle or systemic right ventricle have the worst prognosis. Patients with several forms of congenital heart disease often develop heart failure in their adult years if they have survived childhood, and this is an ever-increasing population as more of these individuals have had successful repair and medical therapy that allows them to survive childhood.

The social/and financial impacts of congenital heart defects are significant. Successful treatment requires highly specialized care. Severe congenital heart disease requires extensive financial resources both in and out of the hospital. Children with developmental delay also require community and school-based resources to achieve optimum functioning. As adults, these children who have survived to adulthood may not be able to work.

Heart Transplantation

Another group of patients worthy of consideration for the Compassionate Allowance list are patients awaiting a heart transplant. These patients must by definition have advanced heart failure and an extremely poor prognosis in order to qualify for a new heart. Patients deemed ill enough for heart transplantation have on average at least a 20% one-year mortality. Patients listed in a transplantation category severe enough to be likely to be offered a heart have at least a 50% one-year mortality. Surprisingly, the mortality rate among heart failure patients is higher than in most patients with cancer. In most cases these patients are unable to drive and require transportation. These patients usually cannot work, so have no income. Some require a constant infusion of an intravenous drug.

For children, 20-25% of those waiting for a transplant do not receive an organ in time due to their severity of illness and the scarcity of donor hearts. The leading causes of heart transplantation in children are age-dependent. The causes are congenital from birth to 1 year. The causes are almost equally split between cardiomyopathy and congenital heart disease between the ages of 1-10 years. For adolescents, cardiomyopathy is the leading indication for heart transplantation.

The Heart Transplant Patient

The heart transplant patient may have a ventricular assist device as a bridge to their new heart, and they often need assistance for wound care and battery change.

It is typically stated that heart transplantation swaps diseases – heart failure goes away, but life as an immunosuppressed host takes over. Rejection, immunosuppression, important comorbidities and malignancies remain obstacles to truly favorable long-term outcomes. Immunosuppression commonly leads to other problems such as kidney dysfunction, hypertension, and peripheral vascular disease.

Some of these patients have significantly impaired exercise capacity despite having nearly normal heart muscle function. This may occur for a variety of reasons. Cardiac rehabilitation is not consistently available after heart transplantation and is not consistently covered by insurance companies. The anti-rejection medications, especially corticosteroids, are prone to induce skeletal muscle weakness which may be further exacerbated by drug-induced diabetes, obesity and hypertension.

The Disability Process for Patients with CVD

Before I conclude my presentation, I would like to address one final subject – the patients I and my fellow health care providers treat who have CVD. It is easy to discuss these conditions in the abstract and explain what symptoms patients with heart failure or congenital heart defects may have, but only by working with and treating these patients, can you begin to understand the challenge these conditions place on their day-to-day living. I've seen patients with CVD change their entire lives to try and cope with the disease: patients who have to get out of bed in stages in order to avoid dizziness when they stand up; patients who get fatigued and short of breath from walking from the front door of their house to their car; patients who can no longer work – who report feeling useless – because they have difficulty walking or standing, or in some cases, even just sitting for substantial periods of time. Young adults may have to rely on their elderly parents

to handle basic chores for them such as bringing in their groceries, because they do not have the strength to do so on their own. Or a young mother may no longer be able to pick up her infant daughter because she isn't allowed to raise her arms over her head. These are the individuals that Social Security disability benefits are meant to help.

But we know the disability application process doesn't always work as intended. Patients often report that the process is too confusing and we can see that it takes too long. I've heard severely impaired patients report that their application was met with skepticism because they didn't look "sick enough". And many patients find that their applications are denied and have to apply again before benefits are ultimately approved. That is why the work the SSA has been doing the past several years – announcing your intent to reevaluate the cardiovascular listings, contracting with the Institute of Medicine to review the current listings, and now today's outreach hearing on Compassionate Allowances – is so important. But we urge the Administration to do more.

In addition to the inclusion of the conditions listed below on the compassionate allowance list, we'd like to see the amount of time between the submission of the application and the approval of disability benefits drop significantly. The Administration could, for example, provide temporary benefits to patients who can provide valid documentation that they have a severe cardiovascular condition. The temporary benefits could remain in place until the SSA was able to conduct a full review of the patient's disability application, or for a set period of time (such as 6 months) before the patient is reevaluated.

We'd also like to reduce the 24-month waiting period patients must currently wait after their disability benefits are approved before becoming eligible for Medicare. Patients who are unable to work because of their CVD are less likely to have health insurance and more likely to need medical care that they are unable to afford.

Consider, for example, John and Sam:

"John" is a 56 year old patient who was diagnosed with advanced heart failure in January 2009 following a heart attack. John was approved for disability in February 2010. Yet because of his lack of health insurance, he hasn't been able to afford the medical therapy that might allow some recovery of his heart function. Instead his condition has worsened and his only option now appears to be a heart transplant. But his Medicare benefits won't begin until 2012 – and he certainly can't afford a transplant without it – though he may not survive long enough to have it.

Or "Sam" a 27 year old who was diagnosed with advanced congestive heart failure in December 2005 and was deemed disabled in July 2008. He had no insurance until he qualified for Medicare this year. This led to limited access to medical care and his condition suffered. Instead of being able to treat him proactively to prevent increased symptoms he would frequently show up at death's door, requiring long hospital admissions. He suffered progressive worsening of symptoms and ultimately cardiogenic shock. By July 2010, when he finally received benefits, he had been hospitalized for a total of 83 days. Over 60 of those days were in the intensive care unit. The total cost, over 1.3 million dollars.

For patients like John and Sam and others like them, the 24-month waiting period is entirely too long and can have devastating effects on their health, as well as significant costs to the health care system. The shame is that these health effects and their costs would often have been preventable.

My most regretful experiences have been in caring for the middle aged adult without Medicaid or Medicare who becomes gravely ill with heart failure and has no option; the older patient and the abject poor are in a better situation under these circumstances.

AHA's Recommendations for the Compassionate Allowance Listing

The ultimate outcome of many cardiovascular diseases and conditions is **heart failure**. When heart failure status and symptoms places patients in NYHA Class III or IV, then disability is a reality.

Because of the medical, physical/emotional, financial, and social limitations imposed by these diagnoses and because of the incurable and disabling course of these diagnoses, the American Heart Association supports the inclusion of the following CVD conditions and circumstances on the compassionate allowance list:

- Advanced HF patients who are candidates for LVAD or heart transplant
- Adults with CVD and class IV symptoms and cyanotic adult congenital heart disease where transplant is the only option
- Class IV symptoms with intractable primary pulmonary hypertension where lung transplantation is the only hope
- Class IV disease where a treatment option exists, (hypertrophic cardiomyopathy [surgery], sarcoid heart disease, amyloidosis, [? immunosuppression], prolonged QT interval with SCD [implanted defibrillator], etc.)
- HF patients, who despite attempts at medical therapy, have 6-min walk distance less than 309 feet, VO2 max less than 50% of predicted, or are dependent upon inotrope therapy or mechanical support
- Fulminant giant cell myocarditis, an imminently fatal disease in which urgent intervention is required
- Lymphomatous invasion of the heart; this is usually from either a primary cardiac tumor [e.g., sarcoma] or contiguous spread from an adjacent lymphoma; these tumors are usually radiation and chemo sensitive but a full assault is required to provide recovery
- Congenital heart disease with Eisenmenger physiology and pulmonary vascular disease
- Congenital heart disease and NYHA Class 4 Symptoms
- Congenital heart disease and severe cyanosis (tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, pulmonary atresia, truncus arteriosus and total anomalous pulmonary venous connection)
- Congenital heart disease after Fontan palliative surgery and protein losing enteropathy
- Congenital heart disease after Fontan, with systolic or diastolic dysfunction
- Transposition of the great vessels after Mustard procedure and RV dysfunction
- Hypoplastic left heart syndrome

- Valvular atresia – aortic, tricuspid, pulmonary, mitral valve
- Single ventricle patients with congestive heart failure
- Childhood myocardial infarction

Conclusion

In closing, I thank you again for the opportunity to present the views of AHA at this meeting and reiterate our appreciation of your willingness to add CVD to the compassionate allowances list as appropriate.

Timely access to Social Security financial resources is critical to survival for many patients with cardiovascular disease. Timely interventions can delay the progression of the disease process and keep patients alive long enough to receive definitive interventions. Having the necessary financial resources afforded by inclusion on the compassionate allowances list ensures that quality of life is optimized in an otherwise desperate situation and is a key factor in reducing morbidity, mortality, and increased medical costs to individuals and to society.

I will be happy to remain available to answer any questions you might have.

AHA's Recommendations for Additions to the Compassionate Allowance List

Diagnosis	Interventions/Measures
Advanced HF patients who are candidates for LVAD or heart transplant	LVAD or transplantation
Adults with CVD and class IV symptoms and cyanotic adult congenital heart disease where transplant is the only option;	Heart Transplantation
Class IV symptoms with intractable primary pulmonary hypertension where lung transplantation is the only hope	Lung Transplantation
Class IV disease where a treatment option exists , (hypertrophic cardiomyopathy [surgery], sarcoid heart disease, amyloidosis, [? immunosuppression], prolonged QT interval with SCD [implanted defibrillator], etc.)	Surgery, Immunosuppression, Pharmacologic agents and ICD
HF patients, who fail medical therapy or are dependent upon inotrope therapy or mechanical support	6-min walk distance less than 309 feet, VO2 max less than 50% of predicted, or dependent upon inotrope therapy or mechanical support
Fulminant giant cell myocarditis , an imminently fatal disease in which urgent intervention is required usually affects young otherwise healthy individuals; rate of death or heart transplantation is approximately 70% at 1 year; post-transplantation survival is approximately 71% at 5 years (PubMed)	Transplantation and immunosuppression
Lymphomatous invasion usually from either a primary cardiac tumor [e.g., sarcoma] or contiguous spread from an adjacent lymphoma; these tumors are usually radiation and chemo sensitive, but a full assault is required to provide recovery of the heart	Cardiac support until recovery
Congenital heart disease with Eisenmenger physiology and pulmonary vascular disease	Surgical correction of cardiac defect and/or treatment of PAH; endocarditis risk
Congenital heart disease and NYHA Class 4 Symptoms	Medical, LVAD or transplantation, social and psychological support.
Congenital heart disease and severe cyanosis (tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, pulmonary atresia, truncus arteriosus and total anomalous pulmonary venous connection)	Surgical repair; endocarditis risk; risk for rhythm abnormalities; lifetime follow-up needed
Congenital heart disease after Fontan palliative surgery and protein losing enteropathy	Heart transplantation; lifetime follow-up needed

Diagnosis	Interventions/Measures
Congenital heart disease after Fontan, with systolic or diastolic dysfunction	Surgical repair vs. heart transplantation; lifetime follow-up
Transposition of the great vessels after Mustard procedure and RV dysfunction	Surgical repair vs. heart transplantation; endocarditis risk; lifetime follow-up
Hypoplastic left heart syndrome	Defect isn't correctable, but some babies can be treated with a series of operations or with a heart transplant; risk for endocarditis; lifelong follow-up required; Virtually all children with HLHS will require heart medicines, heart catheterization and additional surgery.
Valvular atresia – aortic, tricuspid, pulmonary mitral valve	Surgical repair; medications; Risk for endocarditis; Lifelong follow-up required
Single ventricle patients with congestive heart failure Represented by <i>Hypoplastic Left Heart Syndrome (HLHS)</i> ; <i>Pulmonary Atresia/Intact Ventricular Septum</i> ; <i>Tricuspid Atresia</i>	Surgical repair; medications; Risk for endocarditis; Lifelong follow-up required
Childhood myocardial infarction	Multiple causes: common ones include Kawasaki disease and anomalous left coronary artery from the pulmonary artery; medication and surgery (for ex: CABG); severe cases may require heart transplant

AHA's Recommendations for Changes to the Social Security Disability Process

Recommended Change
Reduce the time between submission of disability application and approval of benefits
Offer temporary disability benefits for those with severe CVD until a complete review is completed
Reduce the 24-month Medicare waiting period