

## News Release

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HEALTH  
UK Thalassaemia Society

### Press Conference - Cyprus - Iron Overload

PAPHOS, Cyprus, July 31 /PRNewswire/ --

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TITLE: EXPERTS IN THALASSEMIA CONVENE IN CYPRUS TO DISCUSS REPORTS  
----- OF SIGNIFICANT DECLINES IN HEART DISEASE AND DEATH IN  
PATIENTS WITH THALASSEMIA

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World experts in thalassaemia convened in Paphos to discuss recent findings of decreased heart disease and increased survival in patients with the inherited disease, thalassaemia major. Cyprus has one of the highest recorded carrier rates for this genetic disease, a disorder in which children require blood transfusions every 2-4 weeks to survive. While alleviating their life-threatening anaemia, the iron contained in the transfused red blood cells accumulates over time and deposits in the liver, kidneys, glands and other tissues and, unless an effective iron chelator is administered to these patients, most will die before they reach their teen years.

Over 30 years ago, an injectable iron chelator, deferoxamine, began to be administered to most patients in Cyprus, resulting in a prolongation of life. However, as patients reached their 20's and 30's, heart disease became prevalent and, in spite of deferoxamine therapy, the death rate in Cyprus began climbing until the year 2000, and then the trend reversed. In 2000, the European Union approved Ferriprox(TM) that can be viewed at <http://www.ferriprox.com/Patient/Default.asp>, (deferiprone) an oral iron chelating agent that is used for patients who are inadequately treated with deferoxamine or who have toxicity to it. About one-half of the patients in Cyprus today are treated with deferiprone,

many in combination with deferoxamine. The death rate in thalassemia, which had been climbing between 1980 and 1999, reaching a peak of approximately 6 deaths/1,000 person years between 1995-1999, suddenly dropped to about 2.5 deaths/1,000 person years between 2000-2004, and none of those who died were on deferiprone.

The study reporting this change in mortality in Cypriot thalassaemia patients was reported in July 2006 in *Haematologica*. Two months previously, in the May 2006 issue of *Blood*, Dr. Caterina Borgna-Pignatti and her team reported a similar trend in morbidity and mortality in thalassemia patients in 7 thalassaemia centers in Italy, between January 1995 and December 2003. In a natural history study, they reported that 52 patients treated with deferoxamine developed heart disease, compared to none of the patients treated with deferiprone. Fifteen of these patients died in the deferoxamine-treated group vs none in the deferiprone-treated group.

Two randomized controlled studies published by Dr. Dudley Pennell and his colleagues shed light on one of the primary reasons for these effects. They showed that deferiprone was more effective than deferoxamine in getting iron out of the heart. The first study compared the 2 chelators against each other. The second, which was published in April 2007 in *Circulation*, compared the combination of these chelators against deferoxamine alone. Both studies indicated that the main reason for the decreased heart disease and increased survival is deferiprone's superior ability to remove iron from the heart, decreasing the toxicity of iron in transfused patients with thalassemia. The studies also suggest there may be other factors contributing to the cardio-protective actions of deferiprone, including the prevention of iron toxicity within heart cells, even prior to iron removal, and favourable effects on the major blood vessels, showing an improved endothelial response factor with deferiprone compared to deferoxamine.

The principal investigators of these studies convened in Paphos, Cyprus, along with over 100 other experts in the treatment of thalassemia to discuss these results and other data which can help optimize the treatment of patients with thalassemia.

In addition to the obvious benefits to the patients - less heart disease and increased survival - the healthcare system also benefits when patients are treated with deferiprone because the price of this chelator is equivalent to the first chelator introduced over 30 years ago.

Today, there are 3 iron chelators approved in Europe for treating iron overload in patients with thalassemia: Two of these are manufactured by Novartis in Switzerland, Desferal (deferoxamine), which is administered by nightly infusions, and Exjade (deferasirox), a tablet that is suspended in a drink and swallowed. A third chelator is manufactured by Apotex in Canada, Ferriprox(TM) (deferiprone) and is an oral tablet. The reports of decreased heart disease and increased survival discussed at this conference are related to deferiprone (Ferriprox). Although it is too early to know if there will also be an improvement in heart disease and survival with deferasirox (Exjade), we are hopeful that this will prove to be the case over time. For more information on Ferriprox, please visit:<http://www.ferriprox.com/Patient/Default.asp>.

The Minister of Health of Cyprus opened the Workshop and recognized the contributions that the scientists attending the meeting have made. He noted that there had been a profound impact on decreasing morbidity and mortality in thalassemia patients, a disease that is indigenous to Cyprus.

About UKTS: The U.K. Thalassaemia Society exists to provide support, counseling and information to all those affected by thalassaemia. We co-ordinate and promote research and help educate patients and healthcare professionals on the latest and most successful treatment. We also aim to educate the wider population about thalassaemia and encourage as many people as possible to undergo blood screening so that carriers can be identified and can receive appropriate counseling. Further, we aim to establish links between patients and families and encourage a positive outlook in young thalassaemics to assist them in adhering to their treatment. Our mission statement -UKTS aims to be the definitive source of information, education and research for those affected by, or working with thalassaemian. For

more information, please visit: <http://www.ukts.org/>

About Iron Chelators: Any beneficial medication will also have risks of adverse effects. This is also true for all 3 iron chelators discussed here. To get proper prescribing information, including safety information, please contact the official site of the European Medicines Agency or the respective manufacturers' prescribing information. Examples of links for such information are provided below:

Desferal(TM)(desferrioxamine) Prescribing Information - Novartis:  
[http://www.pharma.us.novartis.com/product/pi.jsp?usertrack.filter\\_applied=true&Novald=7852773765922271376](http://www.pharma.us.novartis.com/product/pi.jsp?usertrack.filter_applied=true&Novald=7852773765922271376)

(Due to the length of this URL, it may be necessary to copy and paste this hyperlink into your Internet browser's URL address field. Remove the space if one exists.)

Exjade(TM)(deferasirox) European Public Assessment Report (EMA):  
<http://www.emea.europa.eu/humandocs/Humans/EPAR/exjade/exjade.htm>

Ferriprox(TM) (deferiprone) (<http://www.ferriprox.com/>)

<http://www.ferriprox.com/Patient/Default.asp>

European Public Assessment Report (EMA):  
<http://www.emea.europa.eu/humandocs/Humans/EPAR/ferriprox/ferriprox.htm>

*Distributed by PR Newswire on behalf of UK Thalassaemia Society*

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