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## Theranexus launches a global offering for an amount of c. 4 million euros and welcomes the American Beyond Batten Disease Foundation as a new shareholder

- *The global offering is composed of a reserved offering for specialized and strategic investors and a public offering for retail investors via the PrimaryBid platform*
- *Issue price of 1.28 euros per new share*
- *Subscription commitments of 2.2 million euros, including 1.15 million euros from the American Beyond Batten Disease Foundation and 1.05 million euros from 4 other investors*
- *Closing of the offering conducted through the PrimaryBid platform on July 11, 2023 at 10 p.m. (CEST) and of the Reserved Offering on July 12, 2023 before market opening (subject to early closing)*

**Lyon, France - July 11, 2023 - 5:40 pm CEST** - Theranexus, an innovative biopharmaceutical company specializing in the treatment of rare neurological diseases (the "**Company**"), today announces the launch of a global offering of approximately 4 million euros (the "**Offering**") through the issuance of new shares at a fixed price of 1.28 euros per share exclusively reserved (i) for specialized and strategic investors falling into the categories below, by way of an accelerated book-building, and (ii) for retail investors through the PrimaryBid platform.

The proceeds of the Global Offering, combined with the Company's existing funds, are intended to contribute to finance the pivotal Phase 3 trial of the Batten-1 drug candidate, designed to treat patients suffering from the rare neurological disease Batten disease. The funds would also be used to finance the other research and development activities of the Company, as well as its general corporate needs.

Theranexus, in partnership with the Beyond Batten Disease Foundation (BBDF), recently obtained approval from the Food and Drug Administration (FDA) and the European Medicines Agency (EMA) for the design of the pivotal phase 3 study of its drug candidate Batten-1. The Company also reports encouraging preliminary results at 6 months from the phase 1/2 study, suggesting a beneficial effect of Batten-1 on neuronal death and stabilization of motor function in treated patients. Encouraged by these latest developments, the Company will launch the pivotal phase 3 of its clinical trial at the end of the second half of 2023. The costs of this phase 3, which should be completed by the end of 2026, are estimated at 5 million euros per year for the next 3 years.

**Mathieu Charvériat, CEO of Theranexus**, said *"We will initiate phase 3 of Batten-1 at the end of 2023, using the funds generated by this operation, and building upon FDA and EMA positive opinions and encouraging preliminary clinical results. The design of the study will enable us to communicate a frequent and rich newsflow to our shareholders, until the product is registered in Europe and the United States. We would like to thank the investors involved in this transaction, and in particular the BBDF Foundation, which, in addition to its strong operational and strategic support for the scientific and clinical development of Batten-1 since 2019, is now committed to supporting us by acquiring a stake in the Company."*

On June 30, 2023, Theranexus had available cash of €4.7 million, reflecting a controlled cash consumption over the second quarter, which included in particular the repayment of the annual loan repayment schedule under the State Guaranteed Loan (PGE) for €0.4 million.

### Global Offering terms and conditions

The Global Offering will be carried out through two distinct but concomitant offers:

- an offer to qualified investors based on article L. 225-138 of the French Commercial Code, via two capital increases:
  - o a first capital increase without shareholders' preferential subscription rights reserved to investors belonging to the category defined in the 18<sup>th</sup> resolution of the Company's annual shareholders' meeting dated June 8, 2023 (the "**AGM**"), i.e. individuals or legal entities (including companies), trusts, investment funds or other investment vehicles of any kind whatsoever, governed by French or foreign law, whether or not they are shareholders in the Company, investing on a regular basis or having invested (including, where applicable, in the form of loans or convertible or non-convertible debt securities) at least two million euros over the 36 months prior to the issue in question in the life sciences or technology sector<sup>1</sup>; and
  - o a second capital increase without shareholders' preferential subscription for investors in the category covered by the 19<sup>th</sup> resolution of the AGM, i.e. the Company's strategic, commercial or financial partners, located in France or abroad, who have entered into or are due to enter into one or more partnership (development, co-development, distribution, manufacturing, etc.), commercial or financing agreements with the Company (or a subsidiary thereto), directly or through one or more entities that these partners control, that control these partners or that are controlled by the same person(s) as these partners, directly or indirectly, within the meaning of Article L. 233-3 of the French Commercial Code<sup>2</sup>;

(the "**Reserved Offering**")

- a capital increase with cancellation of shareholders' preferential subscription rights by way of a public offering (other than an offering covered by 1° of Article L. 411-2 of the French Monetary and Financial Code) primarily aimed at retail investors via the PrimaryBid platform (in accordance with the 15<sup>th</sup> resolution of the General Meeting), with an allocation proportional to demand limited to the amount allocated to this public offering and reduced allocations in the event of excess demand (the "**PrimaryBid Offering**").

The total target amount of the Global Offering would be approximately 4 million euros, including share premium, with the PrimaryBid Offering representing at most 20% of this amount (i.e. 800,000 euros) and not being completed in the absence of the Reserved Offering.

The Reserved Offering will be carried out by accelerated book-building, at the end of which the number of new shares to be issued will be determined. The final size of the Reserved Offering and the PrimaryBid Offering will

<sup>1</sup> i.e. « *les personnes physiques ou morales (en ce compris des sociétés), trusts, fonds d'investissement ou autres véhicules de placement de quelle que forme que ce soit, de droit français ou étranger, actionnaires ou non de la Société, investissant à titre habituel ou ayant investi (y compris, le cas échéant, sous forme de prêt ou de titres de créances convertibles ou non) au moins deux millions d'euros au cours des 36 mois précédant l'émission considérée dans le secteur des sciences de la vie ou des technologies* ».

<sup>2</sup> i.e. « *partenaires stratégiques, commerciaux ou financiers de la Société, situés en France ou à l'étranger, ayant conclu ou devant conclure un ou plusieurs contrats de partenariat (développement, co-développement, distribution, fabrication, etc.), commerciaux ou de financement avec la Société (ou une filiale), directement ou via une ou plusieurs entités que ces partenaires contrôlent, qui contrôlent ces partenaires ou qui sont contrôlés par la ou les mêmes personnes que ces partenaires, directement ou indirectement, au sens de l'article L. 233-3 du code de commerce* ».

depend exclusively on the orders received for each offer, with no reallocation between the PrimaryBid Offering and the Reserved Offering.

The price per share of the Reserved Offering will be €1.28 (representing a discount of 25.8% on the closing price of THERANEXUS shares on July 10, 2023, i.e. €1.73, and a discount of 25.2% to the volume-weighted average price of THERANEXUS shares over the last 3 trading days prior to the pricing of the issue (i.e. July 6, July 7 and July 10, 2023), i.e. €1.71). The subscription price of the new shares offered in the PrimaryBid Offering will be the same as the the subscription price of the new shares offered in the Reserved Offering.

The final number of shares to be issued will be decided by the Company's Chief Executive Officer, by virtue of and within the limits of the sub-delegations of authority granted by the Company's Board of Directors on the date of this press release, it being specified that the maximum number of new shares that may be issued in connection with the Global Offering is 10,000,000 new shares, in accordance with the resolutions of the AGM.

Prior to the launch of the Global Offering, Beyond Batten Disease Foundation, a long-standing partner of the Company and certain financial investors, have undertaken to subscribe to the Reserved Offer for a total amount of 2.2 million euros, representing together approximately 55% of the total amount of the Global Offering. These amounts may be reduced depending on demand.

The table below details the above commitments:

Subscriber's identity	Amount of subscription commitment
Beyond Batten Disease Foundation	€1,150,000
Iris*	€400,000
Gestys*	€300,000
Tocqueville Finance	€250,000
Financière Arbevel	€100,000
<b>Total</b>	<b>€2,200,000</b>

\* In return for their subscription commitment, Iris and Gestys will each receive a guarantee fee equal to 5% of the amount of its commitment (i.e. a fee of 20,000 euros and 15,000 euros respectively), plus, if applicable, an amount equal to 2% of the amount that the concerned investor would actually subscribe for under the Reserved Offer.

The accelerated book-building procedure for the Reserved Offering will be initiated immediately and is expected to close before market opening tomorrow, subject to any early closing. The PrimaryBid Offering will begin immediately and is expected to close at 10 p.m. (Paris time) today, subject to early closing. The Company will announce the results of the Global Offering and, in particular, the number of shares to be issued, as soon as possible after the closing of the order book, through a press release.

The Reserved Offering will be offered, within the categories of investors defined above, (i) to institutional investors in France and outside France, with the exception of the United States, Canada, Australia and Japan, in offshore transactions as defined in and in reliance on Regulation S under the U.S. Securities Act of 1933, as amended (the "Securities Act") and (ii) in a private placement to a limited number of "qualified institutional buyers" within the meaning of Rule 144A under the Securities Act or "institutional accredited investors" within the meaning of Rule 501(a)(1), (2), (3), (7), (8), (9), (12) or (13) of Regulation D under the Securities Act in the United States, pursuant to an exemption from registration under the Securities Act.

Settlement-delivery of the new shares and their admission to trading on the Euronext Growth® Paris multilateral trading facility are expected to take place on July 14, 2023. The new shares will be of the same class and fungible

with the existing shares, will carry all the rights attached to the shares, and will be admitted to trading on the Euronext Growth® Paris multilateral trading facility under the same ISIN code FR0013286259.

The Global Offering is not subject to a prospectus requiring an approval from the French Financial Market Authority (*Autorité des Marchés Financiers*) (the "AMF").

### Abstention and lock-up commitments

As part of the Global Offering, the Company has signed a lock-up commitment that comes into effect on the date of execution of the placement agreement entered into between the Company and Portzamparc today, and continuing for a period of 90 days following the settlement-delivery date of the Global Offering, subject to customary exceptions. The Company's founders, Mr Franck Mouthon, Chairman of the Board of Directors of Theranexus, and Mathieu Charvériat, Chief Executive Officer of Theranexus, and Auriga Partners, an' historical investor of the Company, holding together 22.7% of the Company's share capital, have also signed lock-up commitments taking effect on the date of execution of these commitments and continuing for a period of 180 days following the settlement-delivery date of the Global Offering, subject to certain customary exceptions.

### Financial intermediaries

Portzamparc (BNP Paribas Group) is acting as single global coordinator, lead manager and bookrunner for the Reserved Offering. The Reserved Offering is the subject of a placement agreement between the Company and Portzamparc dated July 11, 2023.

Under the PrimaryBid Offer, investors will only be able to subscribe via the PrimaryBid partners listed on [www.PrimaryBid.fr](http://www.PrimaryBid.fr). The PrimaryBid Offer is the subject of a commitment letter between the Company and PrimaryBid and is not the subject of an investment contract. For further details, please visit [www.PrimaryBid.fr](http://www.PrimaryBid.fr).

### Risk Factors

Your attention is drawn to the risk factors associated to the Company and its business, presented (i) in chapter 3 of the universal 2022 registration document filed with the AMF on April 26, 2023 under number D.23-0345, available free of charge on the Company's website ([www.theranexus.com](http://www.theranexus.com)) and the website of the *Autorité des marchés financiers* ([www.amf-france.org](http://www.amf-france.org)). The occurrence of any or all of these risks could have a material adverse effect on the Company's business, financial condition, results of operations, development or prospects.

Additionally, investors are invited to consider the following risks specific to the issue: (i) the market price of the Company's shares could fluctuate and fall below the subscription price of the shares issued in connection with the Global Offering, (ii) the volatility and liquidity of the Company's shares could fluctuate significantly, (iii) sales of the Company's shares could occur on the market and have an unfavorable impact on the Company's share price, (iv) the Company's shareholders could suffer potentially significant dilution as a result of any future capital increases made necessary by the Company's search for financing, and (v) as the securities are not intended to be listed on a regulated market, investors will not benefit from the guarantees associated with regulated markets.

### About Batten-1

Batten-1 is a new proprietary and exclusive pharmaceutical speciality whose active ingredient is miglustat. The mechanism of action of this active ingredient blocks the accumulation of glycosphingolipids and neuroinflammation, thereby significantly reducing neuronal death, the cause of progressive loss of function in patients. For patients over 17 years of age in the Phase

1/2 study, the product is administered in solid form. In the Phase 3 study, it will be administered in a liquid form, more suited to a paediatric population.

Study design Phase 1/2: This is an open-label study involving 6 patients over 17 years of age with Batten CLN3 disease, treated with miglustat up to 600 mg/day for 2 years. The primary endpoint was safety and tolerability, assessed by adverse event reporting, bioassays and ECG, as well as miglustat pharmacokinetics. Secondary endpoints included efficacy parameters: the Unified Batten Disease Rating Scale, visual acuity, measurement of brain volumes by magnetic resonance imaging and measurement of retinal neuronal layer thickness by optical coherence tomography. Batten-1 dose escalation to a maximum of 600 mg/d was well tolerated, with no serious adverse events or discontinuations. The most frequently reported adverse events were gastrointestinal, reversible and mostly mild to moderate in intensity, demonstrating the good safety profile of Batten-1 in this population. Batten-1 will continue to be evaluated in these patients for a total of 24 months. Further information is available at <https://clinicaltrials.gov/ct2/show/NCT05174039>.

## About Batten disease

Juvenile Batten disease or Spielmeyer-Vogt disease, also known as CLN3 disease, is a rare, fatal genetic disorder of the nervous system for which there is no cure. It belongs to the group of neuronal ceroid-lipofuscinoses (NLC). Over 400 mutations in 13 genes have been associated with NLC variants, which differ essentially in the age of onset of symptoms. The first symptom of the juvenile form, progressive vision loss, appears between the ages of 4 and 6. This is followed by cognitive and behavioral problems and motor difficulties. Convulsions appear within 2 to 4 years of onset. Gradually, patients decline both physically and mentally. Eventually, they become wheelchair-bound or even bedridden, before dying prematurely.

The disease is always fatal - usually in late adolescence or early adulthood. In the USA and Europe, Batten disease is the most prevalent NLC, affecting around 2,000 patients<sup>3</sup>. From a pathophysiological point of view, interactions between neurons and glial cells play a key role in the onset and progression of all NLCs.

## About the Beyond Batten Disease Foundation (BBDF)

The Beyond Batten Disease Foundation (BBDF) is the world's largest non-profit organization dedicated to funding research to treat and cure juvenile Batten disease (CLN3). Since its creation in 2008, over \$35 million has been invested in research through donations, co-funding and strategic partnerships. BBDF spearheads a unique and coherent strategy, integrating independent scientific resources and collaboration with related organizations to conduct research into juvenile Batten disease. BBDF-sponsored research initially uncovered the mechanisms of the disease<sup>4</sup>. Today, a treatment is in sight. BBDF-funded research has led to the discovery of a drug, Batten-1, which slows disease progression in models of Batten disease. For more information, visit [www.beyondbatten.org](http://www.beyondbatten.org).

## About Theranexus

Theranexus is an innovative biopharmaceutical company spun out of the French Atomic Energy Commission (CEA), specializing in the treatment of rare neurological disorders. The company has a unique platform for the identification and characterization of innovative therapy drug candidates in the field of rare neurological disorders, and a first drug candidate in development and in the clinic for Batten disease.

THERANEXUS is listed on Euronext Growth in Paris (FR0013286259- ALTHX).

More information at

<http://www.theranexus.com>

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## Contact

<sup>3</sup> National Organization for Rare Disorders (NORD)/Orphanet

<sup>4</sup> Settembre et al, TFEB links autophagy to lysosomal biogenesis, Science 2011  
EUI-1216371163v1

**THERANEXUS**

**Thierry LAMBERT**  
Chief Financial Officer  
[contact@theranexus.com](mailto:contact@theranexus.com)

**NEWCAP**

**Théo MARTIN/Pierre LAURENT**  
Investor Relations  
+33 (0)1 44 71 94 97  
[theranexus@newcap.eu](mailto:theranexus@newcap.eu)

**FP2COM**

**Florence PORTEJOIE**  
Media Relations  
+ 33 (0)6 07 76 82 83  
[fportejoie@fp2com.fr](mailto:fportejoie@fp2com.fr)

**Disclaimer**

In France, the offer of Theranexus shares described below will be carried out through (i) a reserved offering and (ii) a public offer primarily intended to retail investors through the PrimaryBid platform. Pursuant to article 211-3 of the General regulations of the French financial markets authority (*Autorité des marchés financiers*) (the "**AMF**") and articles 1(4) and 3 of the Regulation (EU) 2017/1129 of the European Parliament and of the Council of 14 June 2017, as amended (the "**Prospectus Regulation**"), the offer of Theranexus shares will not require the publication of a prospectus approved by the AMF.

With respect to Member States of the European Economic Area, no action has been taken or will be taken to permit a public offering of the securities referred to in this press release requiring the publication of a prospectus in any Member State. Therefore, such securities may not be and shall not be offered in any Member State other than in accordance with the exemptions of Article 1(4) of Prospectus Regulation or, otherwise, in cases not requiring the publication of a prospectus under Article 3 of the Prospectus Regulation and/or the applicable regulations in such Member State.

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