ASX/NASDAQ ANNOUNCEMENT

Benitec receives U.S. Orphan Drug Designation for BB-301, its ddRNAi therapeutic for the treatment of oculopharyngeal muscular dystrophy

---

Sydney, Australia, 15 January 2018: Benitec Biopharma Limited (ASX:BLT; NASDAQ:BNTC; NASDAQ:BNTCW) today announced that the U.S. Food & Drug Administration (FDA) has granted Orphan Drug Designation to BB-301 for the treatment of oculopharyngeal muscular dystrophy (OPMD).

The Orphan Drug Designation granted to Benitec may provide a range of valuable benefits, including fast track process for clinical regulatory approval, potential extension of patent life with a seven-year period of market exclusivity if the drug is approved, tax credits for qualified clinical trials and an exemption from FDA application fees. In short, a clear and expeditious path for cost-efficient development and commercialisation. The granting of orphan status from the FDA in the US follows on from receiving orphan designation from the European Medicines Agency in early 2017.

Greg West, CEO, Benitec Biopharma, commented on today's news, "We are very pleased to have received Orphan Drug Designation from the FDA for BB-301, as it is another significant step forward for a key program in our pipeline. We believe BB-301 represents a promising new approach for the treatment of OPMD and has the potential to make a meaningful impact for patients who have this debilitating disease. The Benitec team is focused on executing our plan to advance BB-301 into human clinical trials by the end of 2018."

BB-301 is a single vector (gene therapy construct) system which uses DNA directed RNA interference (ddRNAi) to silence expression of the mutant gene associated with OPMD, while simultaneously adding back a copy of the normal version of the same gene to restore gene function. Nonclinical safety studies and manufacturing work are progressing and Benitec intends to file an Investigational New Drug Application (IND) in the last quarter of calendar year 2018.

In November last year Benitec advised it had completed pre-investigational new drug application (pre-IND) and scientific advice meetings with the U.S. FDA, Health Canada and several European agencies. The purpose of these meetings was to discuss the regulatory development pathway for BB-301 as a treatment for OPMD and to ensure Benitec's proposed development program addressed the regulatory expectations of these agencies. In addition to these regulatory meetings, the transfer of production protocols and optimisation of the processes related to manufacturing of BB-301 are well underway at Benitec's contract manufacturing organisation (CMO).

Mr. West concluded, “We also believe this program, if successful, can act as a proof of concept for using our groundbreaking ‘silence and replace’ technology for other therapeutic targets potentially expanding market opportunities for Benitec and paving the way for the development of other monogenic orphan disease programs in the future. Management is dedicated to generating shareholder value through this and our other promising programs currently in the development pipeline, and I look forward to sharing 2018 updates with the market as they occur.”

The FDA's Office of Orphan Drug Products may grant orphan status to support development of medicines for the treatment of rare diseases that affect fewer than 200,000 people in the United States.

For further information regarding Benitec and its activities, please contact the persons below, or visit the Benitec website at www.benitec.com

Australia Investor Relations

Market Eye
Orla Keegan
Director
Tel: +61 (2) 8097 1201
Email: orla.keegan@marketeye.com.au

United States Investor Relations

M Group Strategic Communications
Jay Morakis
Managing Director
Tel: +1 212.266.0191
Email: jmorakis@MGroupSC.com

About Benitec Biopharma Limited:

Benitec Biopharma Limited (ASX: BLT; NASDAQ: BNTC; NASDAQ: BNTCW) is a biotechnology company developing innovative therapeutics based on its patented gene-silencing technology called ddRNAi or 'expressed RNAi'. Based in Sydney, Australia with laboratories in Hayward, California (USA), and collaborators and licensees around the world, the company is developing ddRNAi-based therapeutics for chronic and life-threatening human conditions including head & neck squamous cell carcinoma, OPMD retinal based diseases such as wet age-related macular degeneration, and hepatitis B. Benitec has also licensed ddRNAi to other biopharmaceutical companies for applications including HIV/AIDS, Huntington's Disease, chronic neuropathic pain, cancer immunotherapy and retinitis pigmentosa.

About OPMD:

OPMD is a rare inherited myopathy characterized by dysphagia (difficulty in swallowing), the loss of muscle strength, and weakness in multiple parts of the body. Patients typically suffer from severe dysphagia, ptosis (eye lid drooping), tongue atrophy, proximal lower limb weakness, dysphonia (altered and weak voice), limitation in looking upward, as well as facial muscle and proximal upper limb weakness. Progressing throughout that patient's life, OPMD is not typically diagnosed until the individuals reach their 50's or 60's. As the dysphagia becomes more severe, patients become malnourished, lose significant weight, become dehydrated and suffer from repeated incidents of aspiration pneumonia. The last two symptoms are often the cause of death. No cure is currently available for OPMD. The cricopharyngeal myotomy is the only treatment available to improve swallowing in these patients, but because the root cause of the genetic disease has not been addressed, the pharyngeal musculature still undergoes progressive degradation leading to the previously mentioned complications.

Safe Harbor Statement:

This press release contains "forward-looking statements" within the meaning of section 27A of the US Securities Act of 1933 and section 21E of the US Securities Exchange Act of 1934. Any forward-looking statements that may be in this ASX/Nasdaq announcement are subject to risks and uncertainties relating to the difficulties in Benitec's plans to develop and commercialize its product candidates, the timing of the initiation and completion of preclinical and clinical trials, the timing of patient enrolment and dosing in clinical trials, the timing of expected regulatory filings, the clinical utility and potential attributes and benefits of ddRNAi and Benitec's product candidates, potential future out-licenses and collaborations, the intellectual property position and the ability to procure additional sources of financing. Accordingly, you should not rely on those forward-looking statements as a prediction of actual future results.