MEDIA RELEASE. UNDER EMBARGO UNTIL 1 AUGUST, 2020.

New medicine listed on the PBS for the treatment of Dravet syndrome, a rare and refractory childhood-onset epilepsy[1]

Melbourne, Australia. 1 August, 2020 - Diacomit® (stiripentol), the first new treatment for seizures associated with Dravet syndrome in over a decade[2], will be available on the Pharmaceutical Benefits Scheme (PBS) from August 1, 2020.

This listing follows the Therapeutic Goods Administration (TGA) approval of Diacomit® in 2019, based on positive results from two pivotal, double-blind, randomised, placebo-controlled trials.[3] In both trials, Diacomit® in combination with valproate and clobazam, achieved a significantly higher response rate compared to placebo, where a response was defined as a patient who achieved ≥50% decrease in the frequency of generalised clonic or tonic-clonic seizures during the double-blind treatment period compared to baseline ($p<0.0001$).[4,5]

Dravet syndrome, previously known as severe myoclonic epilepsy of infancy (SMEI), is a rare and refractory form of epilepsy presenting in the first year of life.[6] Children and adults with Dravet syndrome experience severe symptoms, including repeated and prolonged seizures, cognitive and psychomotor impairment, as well as developmental delay.[7]

Professor Ingrid Scheffer, paediatric neurologist and epileptologist, and clinical researcher at Austin Health and the University of Melbourne said:

“Diacomit is an important addition to our treatment options for Dravet syndrome, which is a devastating form of epilepsy. Diacomit is particularly useful for reducing episodes of status epilepticus, where prolonged life-threatening seizures occur; these are a frequent feature in Dravet syndrome.”

Graeme Shears, CEO Epilepsy Foundation, welcomed the new treatment option and improved access for Australians living with epilepsy.

“The treatment of Dravet syndrome has been relatively unchanged for the past decade,” said Mr Shears.“We are pleased that in more recent years we are seeing developments for the treatment of Dravet syndrome, thereby providing patients and their families with greater treatment options.”

Diacomit® marks the first medicine to become available on the PBS under the newly announced organisation, Chiesi Australia.

“The availability of Diacomit® on the PBS is an important step forward for the management of childhood epilepsy, and will lead to improved access for those living with Dravet syndrome, providing them with greater treatment options in an area where there are limited therapeutic opportunities” said **Chris Rossidis, General Manager, Chiesi Australia.**

References

1. Stiripentol Public Summary Document, March 2020 PBAC Meeting, available at: https://www.pbs.gov.au/industry/listing/elements/pbac-meetings/ psd/2020-03/files/stiripentol-psd-march-2020.pdf
2. Epilim Product Information 1991
3. Diacomit Product Information 2020
4. Chiron C, Stiripentol in severe myoclonic epilepsy in infancy: a randomised placebo-controlled syndrome-dedicated trial. *Lancet* 2000;356:1638-42
5. Guerrini R, Stiripentol in severe myoclonic epilepsy in infancy (SMEI): a placbo-controlled Italian trial. *Epilepsia* 2002:43(8):155
6. Dravet C, The core Dravet syndrome phenotype *Epilepsia* 2011;52(2):3-9
7. Guzzetta F, Cognitive and behavioral characteristics of children with Dravet syndrome: An overview *Epilepsia,* 2011;52(2):35-38

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PBS Information: capsules and powder for oral suspension: Authority Required (Streamlined). Refer to PBS schedule for full authority information.

Please review the Product Information before prescribing, available from Chiesi Australia Medical Information (03) 9077 4486.

DIACOMIT Minimum Product Information. Indication: adjunctive treatment of generalised tonic-clonic and clonic seizures associated with severe myoclonic epilepsy in infancy (SMEI, also known as Dravet syndrome) in patients whose seizures are not adequately controlled with a benzodiazepine (usually clobazam) and valproate. **Dosage and administration:** treatment should be initiated by a neurologist experienced in the diagnosis and management of epilepsy. **Recommended dosage:** 50 mg/kg/day, in two to three divided doses. **Dose escalation**: gradual, starting with 20 mg/kg/day for 1 week, then 30 mg/kg/day for 1 week. Further dosage escalation is age dependent: children < 6 years should receive an additional 20 mg/kg/day in the third week, thus achieving the recommended dose of 50 mg/kg/day in three weeks; children 6 - 12 years should receive an additional 10 mg/kg/day each week, thus achieving the recommended dose of 50 mg/kg/day in four weeks; children and adolescents ≥12 years should receive an additional 5 mg/kg/day each week until the optimum dose is reached based on clinical judgment. **Method of administration:** capsules should be swallowed whole and powder mixed in a glass of water and taken immediately; always take with food, avoid milk/dairy products, carbonated drinks, fruit juice, food and drinks containing caffeine or theophylline. **Contraindications:** Hypersensitivity to active ingredient or to any of the excipients; history of psychoses in the form of episodes of delirium. **Precautions:** children <3 years, adults, and elderly; pregnancy category B3; hepatic and renal impairment; driving or operating machinery; monitor liver function and blood count prior to starting treatment and every 6 months; suicidal ideation and behaviour; somnolence and drowsiness, when combined with other central nervous depressants. Dosage adjustment of concomitant clobazam or other anti-epileptic drugs could be considered. All patients treated with antiepileptic drugs, irrespective of indication, should be monitored for signs of suicidal ideation and behaviour, appropriate treatment should be considered at the onset of side effects. **Interactions:** benzodiazepines, non-benzodiazepines, barbiturates, bromides, neuroactive steroids; statins; immunosuppressants; substances metabolised by CYP2C19 and CYP3A4 e.g. citalopram, omeprazole, HIV drugs, antihistamines, calcium channel blockers, statins, oral contraceptives, codeine; carbamazepine,

phenytoin, and phenobarbital have the potential to worsen seizure activity and should not be used with stiripentol in the management of Dravet syndrome; daily dosage of clobazam and/or valproate should be reduced according to the onset of side effects whilst on stiripentol therapy. **Adverse events:** *Very common*: Anorexia, loss of appetite, weight loss, insomnia, drowsiness, ataxia, hypotonia, dystonia. *Common*: neutropenia, persistent severe neutropenia, aggressiveness, irritability, behaviour disorders, opposing behaviour, hyperexcitability, sleep disorders, hyperkinesias, nausea, vomiting, raised γGT. Date of first approval: 13 September 2019. Date of most recent amendment: 28 February 2020.

DIACOMIT® is a registered trademark of Biorganon SA Switzerland. Distributed in Australia by Emerge Health Pty Ltd, a Chiesi Company. Suite 3, 22 Gillman Street, Hawthorn East, VIC. 3123, Australia. Tel: +61 3 9077 4486; Fax: +61 3 8672 0792; Email: customerservice.au@chiesi.com; Website: www.chiesi.com.au. Date of preparation: July 2020.

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About Chiesi Australia

Chiesi Australia is the Australia-New Zealand division of the Chiesi Group, a global pharmaceutical company based in Parma, Italy. The Chiesi Group acquired Emerge Health in November 2019. The Chiesi Group (also known as Chiesi Farmaceutica) is an international research focused Group with over 80 years' experience in healthcare, operating in 29 countries. The Group conducts research, develops treatment options and supplies innovative drugs to treat a wide range of conditions. The Group's Research and Development Centre is based in Parma, Italy and coordinates the activities of four important R&D groups in France, USA, UK and Sweden to drive its own pre-clinical, clinical and registration programs. The Group continues to focus in areas of respiratory diseases, specialty care and rare diseases.