



FOR IMMEDIATE RELEASE:
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**ONY INC., A PIONEER IN NEONATAL PHARMACEUTICAL DEVELOPMENT,
HAS ENTERED INTO A CO-PROMOTION AGREEMENT WITH RECORDATI
RARE DISEASES FOR NEOPROFEN®**

Amherst, N.Y. - ONY, Inc., a pioneer in neonatal pharmaceutical development, has reached an agreement with Recordati Rare Diseases to co-promote NeoProfen® (ibuprofen lysine) Injection.

NeoProfen® is indicated to close a clinically significant Patent Ductus Arteriosus (PDA) in premature infants weighing between 500 and 1500 grams, who are no more than 32 weeks gestational age when usual medical management (e.g., fluid restriction, diuretics, respiratory support, etc.) is ineffective.

ONY, is the manufacturer and marketer of Infasurf® (calfactant) a lung surfactant for the prevention and treatment of Respiratory Distress Syndrome (RDS) in premature infants.

"The co-promotion provides ONY a second product used exclusively in Neonatal Intensive Care Units, and continues the company's commitment to neonatology," said John Librie, CEO, ONY. "Recordati Rare Diseases is an excellent partner that shares the priorities of ONY and whose expertise is complementary to ONY's strengths."

"We have evaluated all of the scientific and medical evidence available for NeoProfen® and believe it is an excellent and important neonatology product for the treatment of Patent Ductus Arteriosus. We are proud to be able to represent NeoProfen® to those caring for premature babies with Patent Ductus Arteriosus" said Edmund A. Egan, M.D., ONY Chairman and Chief Medical Officer.

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About ONY:

ONY Inc. is a small, privately held, biopharmaceutical company focused on the research, development and commercialization of products for the hospital and acute care markets.

ONY was founded by Dr. Edmund Egan, Professor of Pediatrics, State University of New York at Buffalo. Dr. Egan is a pioneering neonatologist whose team conducted trials of one of just three FDA-approved drugs to treat lungs of premature babies at Buffalo Women & Children's Hospital.

ONY is located in the Baird Research Park in Amherst, N.Y.

About Recordati Rare Diseases:

Recordati Rare Diseases (RRD) Inc. began marketing products to treat rare diseases in 2013. RRD is a member of the Recordati Group, which consists of Recordati S.p.A. and Orphan Europe. The Recordati Group was established in 1926 and now has over 3,200 employees worldwide.

RRD's mission is to partner with patients, health care providers, advocacy and industry to make products available to treat rare and severe diseases.

RRD's product portfolio consists of:

- Carbaglu® (carglumic acid)
- NeoProfen® (ibuprofen lysine) Injection
- Panhematin® (hemin for injection)

Other products distributed by RRD are Chemet® (succimer capsules), Cosmegen® (dactinomycin for injection), Desoxyn® CII (methamphetamine hydrochloride tablets, USP), Mustargen® (mechlorethamine HCl for injection), Peganone® (ethotoin tablets, USP), and Tranxene® CIV (clorazepate dipotassium).

About NeoProfen:

NeoProfen is indicated to close a clinically significant Patent Ductus Arteriosus (PDA) in premature infants weighing between 500 and 1500 grams, who are no more than 32 weeks gestational age when usual medical management is ineffective. The clinical trial was conducted among infants with an asymptomatic PDA. However, the consequences beyond 8 weeks after treatment have not been evaluated; therefore, treatment should be reserved for infants with clear evidence of a clinically significant PDA.

NeoProfen is contraindicated in preterm infants with: proven or suspected infection that is untreated; congenital heart disease in whom patency of the PDA is necessary for satisfactory pulmonary or systemic blood flow (e.g., pulmonary atresia, severe tetralogy of Fallot, severe coarctation of the aorta); who are bleeding, especially those with active intracranial hemorrhage or gastrointestinal bleeding; with thrombocytopenia; with coagulation defects; with or who are suspected of having necrotizing enterocolitis; with significant impairment of renal function.

There are no long-term evaluations of the infants treated with ibuprofen at durations greater than the 36 weeks post-conceptual age observation period. Ibuprofen's effects on neurodevelopmental outcome and growth as well as disease processes associated with prematurity (such as retinopathy of prematurity and chronic lung disease) have not been assessed.

NeoProfen may alter the usual signs of infection. The physician must be continually on the alert and should use the drug with extra care in the presence of controlled infection and in infants at risk of infection.

NeoProfen, like other non-steroidal anti-inflammatory agents, can inhibit platelet aggregation. Preterm infants should be observed for signs of bleeding.

Ibuprofen has been shown to displace bilirubin from albumin binding-sites; therefore, it should be used with caution in patients with elevated total bilirubin.

NeoProfen should be administered carefully to avoid extravascular injection or leakage, as solution may be irritating to tissue.

Most common adverse reactions ($\geq 10\%$) are sepsis, anemia, intraventricular bleeding, apnea, gastrointestinal disorders, impaired renal function, respiratory infection, skin lesions, hypoglycemia, hypocalcemia, and respiratory failure.

About Infasurf:

Infasurf is indicated for the prevention of Respiratory Distress Syndrome (RDS) in premature infants at high risk for RDS and for the treatment of premature infants who develop RDS. Infasurf decreases the incidence of RDS, mortality due to RDS, and air leaks associated with RDS.

Prophylaxis

Prophylaxis therapy at birth with Infasurf is indicated for premature infants <29 weeks of gestational age at significant risk for RDS. Infasurf prophylaxis should be administered as soon as possible, preferably within 30 minutes after birth.

Treatment

Infasurf therapy is indicated for infants 72 hours of age with RDS (confirmed by clinical and radiologic findings) and requiring endotracheal intubation.

Important Safety Information

Infasurf is intended for intratracheal use only. THE ADMINISTRATION OF EXOGENOUS SURFACTANTS, INCLUDING INFASURF, OFTEN RAPIDLY IMPROVES OXYGENATION AND LUNG COMPLIANCE. Following administration of Infasurf, patients should be carefully monitored so that oxygen therapy and ventilatory support can be modified in response to changes in respiratory status.

Infasurf therapy is not a substitute for neonatal intensive care. Optimal care of premature infants at risk for RDS and new born infants with RDS who need endotracheal intubation requires an acute care unit organized, staffed, equipped, and experienced with intubation, ventilator management, and general care of these patients.

TRANSIENT EPISODES OF REFLUX OF INFASURF INTO THE ENDOTRACHEAL TUBE, CYANOSIS, BRADYCARDIA, OR AIRWAY OBSTRUCTION HAVE OCCURRED DURING THE DOSING PROCEDURES that required stopping Infasurf and taking appropriate measures to alleviate the condition. After the patient is stable, dosing can proceed with appropriate monitoring.

An increased proportion of patients with both intraventricular hemorrhage (IVH) and periventricular leukomalacia (PVL) was observed in Infasurf-treated infants in the Infasurf- Exosurf Neonatal controlled trials. These observations were not associated with increased mortality.

The most common adverse reactions associated with Infasurf dosing procedures in the controlled trials were cyanosis (65 percent), airway obstruction (39 percent), bradycardia (34 percent), reflux of surfactant into the endotracheal tube (21 percent), requirement for manual ventilation (16 percent), and reintubation (3 percent). These events were generally transient and not associated with serious complications or death.

The incidence of common complications of prematurity and RDS in the four controlled Infasurf trials are presented in the attached table. Prophylaxis and treatment study results for each surfactant are combined.

Common Complications of Prematurity and RDS in Controlled Trials	INFASURF (n=1001), %	Exosurf Neonatal® (n=978), %	INFASURF (n=553), %	Survanta® (n=566), %
Apnea	61	61	76	76
Patent ductus arteriosus	47	48	45	48
Intracranial hemorrhage	29	31	36	36
Severe intracranial hemorrhage	12	10	9	7
IVH and PVLb	7	3	5	5
Sepsis	20	22	28	27
Pulmonary air leaks	12	22	15	15
Pulmonary interstitial emphysema	7	17	10	10
Pulmonary hemorrhage	7	7	7	6
Necrotizing enterocolitis	5	5	17	18
a Grade III and IV by the method of Papile. b Patients with both intraventricular hemorrhage and periventricular leukomalacia.				

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