



Corporate Presentation

March 2021



TARA-002 for Lymphatic Malformations - An investigational broad immunostimulatory cellular therapy for the treatment of Lymphatic Malformations.



IV Choline Chloride for IFALD - An investigational phospholipid substrate replacement therapy for intestinal



TARA-002 for Lymphatic Malformations - An investigational broad immunostimulatory cellular therapy for the treatment of Lymphatic Malformations.

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Modernizing and Expediting Development of De-Risked Assets



Lead programs: TARA-002 in Non-Muscle Invasive Bladder Cancer (NMIBC) and Lymphatic Malformations (LMs)

- Cell-based immunopotentiator
- FDA confirmed initial comparability to Japanese predecessor OK-432
- NMIBC clinical development plan in place following Pre-Investigational New Drug (PIND) engagement with FDA
- TARA-002/OK-432 is standard of care in Japan for LMs; completed Phase 2 study in U.S. supports treatment effect with support for strong safety profile



Other mid-stage development programs provide diversification and additional growth potential

- IV Choline in intestinal failure associated liver disease (IFALD): Completed End Of Phase 2 dialogue with FDA and aligned on Phase 3 design



Company well funded through anticipated key milestones through early 2023

Pipeline Addresses Multiple Indications With High Unmet Need

	PRE-IND	Phase 1	Phase 2	Phase 3
IMMUNOLOGY, ONCOLOGY				
TARA-002 – Lyophilized, inactivated Group A <i>Streptococcus</i>				
Lymphatic Malformations (LMs)*				
Non-Muscle Invasive Bladder Cancer (NMIBC)				
HEPATOLOGY, GI, METABOLICS				
IV Choline Chloride for Injection – Phospholipid Substrate Replacement				
Intestinal Failure Associated Liver Disease (IFALD)**,†				
OTHER				
Vonapanitase – Recombinant Human Type 1 Elastase (phase 1 studies completed in fistula patency and PAD)				

TARA-002

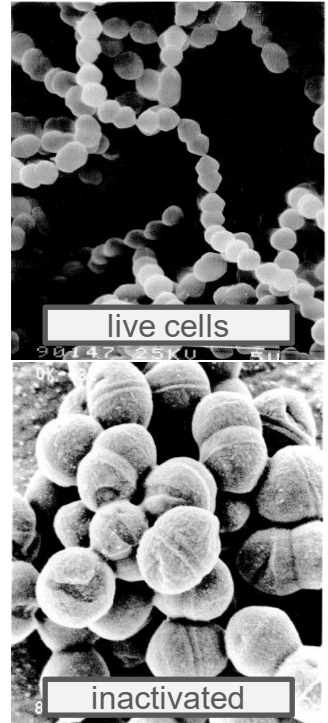
LYOPHILIZED, INACTIVATED GROUP A
STREPTOCOCCUS PYOGENES



...d immunostimulatory cellular therapy
...nephatic Malformations.
Folate Met MAT1A MAT2A
 $H_3C-N^+(CH_3)_2-CH_2OH$
THF MTHFR
CHOLINE
A KEY FACTOR
essential nutrient
- An
substrate
inal
(LD)
of
Streptococcus pyogenes
Choline Chloride for IFALD - An
investigational phospholipid substrate
replacement therapy for intestinal
failure-associated liver disease (IFALD)

TARA-002: Cell-Based Immunopotentiator with Significant Potential

- TARA-002 is an investigational, genetically distinct strain of *Streptococcus pyogenes* that is inactivated while retaining its immune-stimulating properties
- TARA-002 is manufactured under GMP conditions from the same Master Cell Bank as OK-432⁽¹⁾, once one of the largest selling oncology products in Japan
- FDA has confirmed initial comparability between TARA-002 and OK-432 and path forward to completion of GMP comparability
- Having established initial comparability to OK-432, the extensive data generated by OK-432 will help support TARA-002

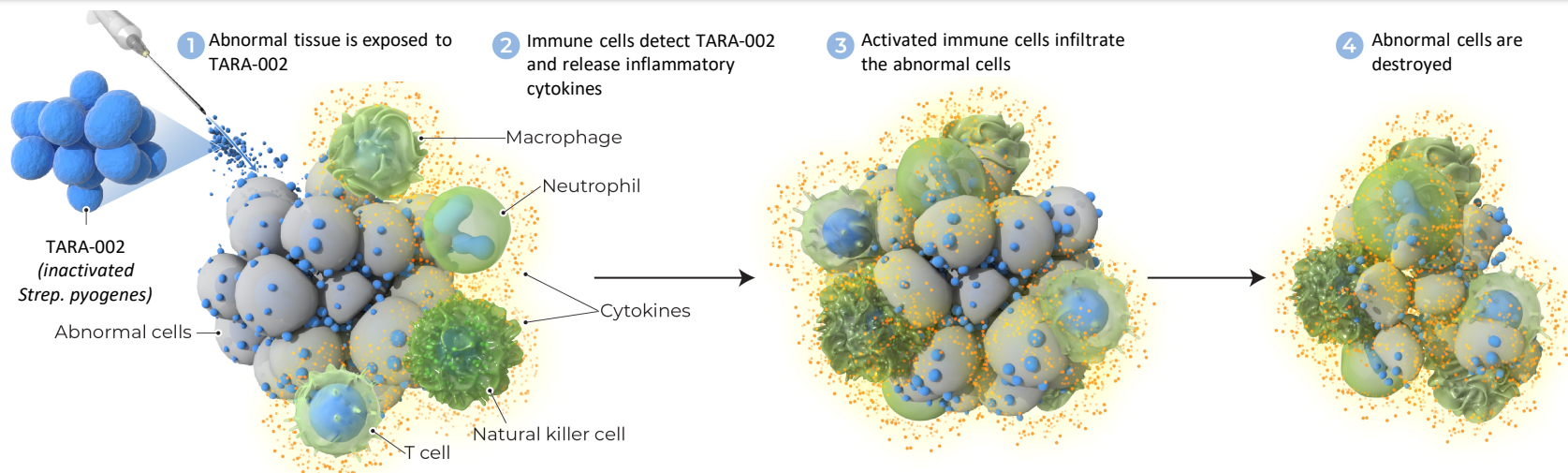


TARA-002: Mechanism of Anti-Tumor / Anti-Cystic Activity

Th1 Like Anti-Tumor Cytokine Response

Multi-Cytokine Inducer⁽¹⁾⁽²⁾⁽³⁾

IL-2 IL-6 IL-8 IL-10 IL-12 GM-CSF G-CSF TNF- α IFN- γ



OK-432: Human Efficacy Data in Multiple Indications

OK-432 has been approved (ex-US) or studied in multiple indications

APPROVED INDICATIONS IN JAPAN⁽¹⁾



- **Lymphangiomas (Lymphatic Malformations)**
- Gastric cancer combo with chemo (post-operative)
- Primary lung cancer combo with chemo
- Reduction of ascites in gastrointestinal cancer
- Reduction of pleural effusion in lung cancer
- Unresponsive head, neck & thyroid cancer

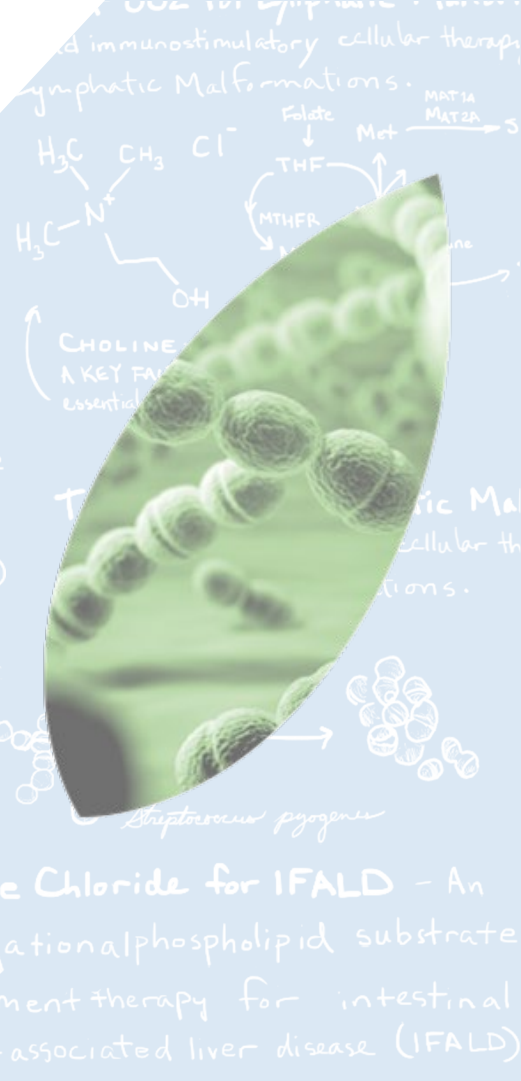


OK-432 CLINICAL RESEARCH CONDUCTED IN:

- **Non-Muscle Invasive Bladder Cancer**
- Ovarian cancer
- Malignant mesothelioma
- Pancreatic cancer
- Esophageal cancer
- Oral squamous cell cancer
- Hepatocellular cancer
- Ranula
- Thyroglossal cysts
- Pleurodesis
- Seroma
- Symptomatic lymphocele
- Auricular hematoma

TARA-002

NON-MUSCLE INVASIVE BLADDER CANCER (NMIBC)

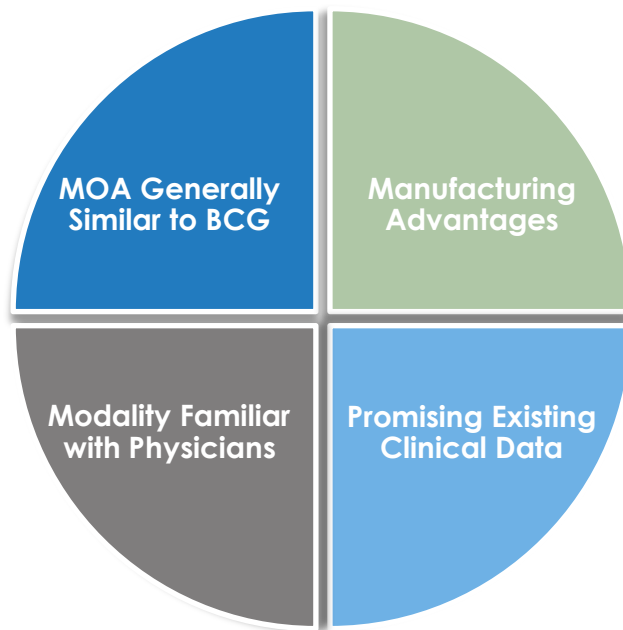


TARA-002 in NMIBC: Profile Supports Potential in NMIBC

Similar mechanism to BCG, notable patient experience in Asia and manufacturing advantages

- Prompts a predominantly Th1 type cytokine response
- Mechanistically similar to Bacille Calmette-Guérin (BCG)

- Urologists have been using an attenuated bacteria, BCG, as immunotherapy for decades
- Intravesical administration is preferred clinical approach among urologists⁽¹⁾



- State-of-the-art U.S. manufacturing facility
- TARA-002 manufacturing process supported by 40 years of production history of OK-432
- ~150 NMIBC patients tested with OK-432 demonstrated promising results⁽²⁾
- Treatment generally well tolerated

Clinical Evidence of OK-432 Provides Strong Rationale for Development of TARA-002 in NMIBC

Data across multiple investigator-initiated studies in ~150 NMIBC patients treated intravesically shows that OK-432:

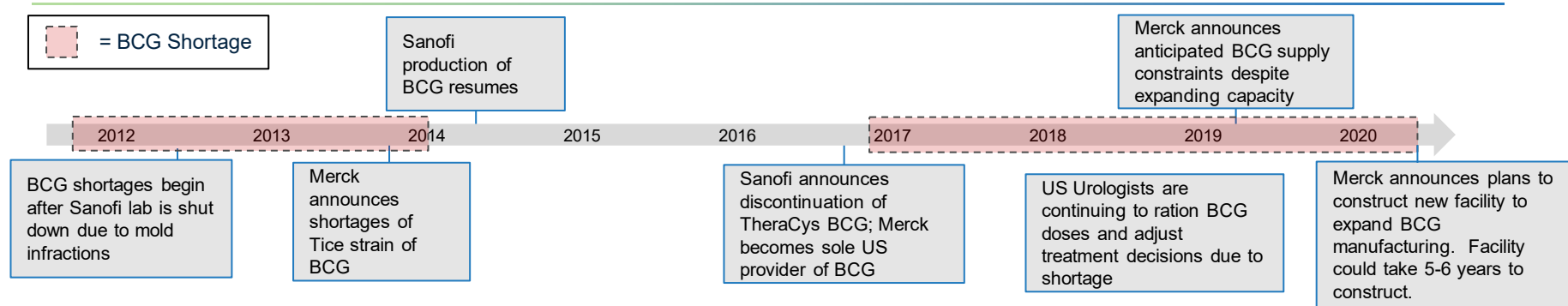


Was generally well-tolerated, with safety and tolerability observed across a range of doses



Demonstrated treatment effect and lower rates of recurrence vs. control group, including in the randomized, controlled setting

BCG Shortage Causes Significant Impact on Care



Shortage has prompted major urological associations to issue guidance on patient management ⁽¹⁾

“...remain extremely concerned about this shortage and its effects on the care of bladder cancer patients...”

-Joint Statement on BCG Shortage, Feb 2019



American
Urological
Association



LUGPA
Integrated Practices
Comprehensive Care



Urology Care
FOUNDATION™
The Official Foundation of the
American Urological Association



Dose rationing and resorting to less desirable treatment options are impacting patient care ⁽²⁾

*“I see patients **every week** whose **treatment decisions** are affected by **the BCG shortage**...sometimes I just recommend moving patients more quickly to cystectomy because **we don’t have any better options available.**”*

-Academic Hospital Urologist

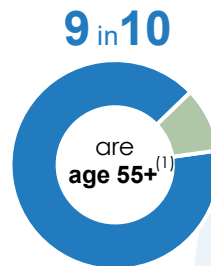
- (1) AUA/SUO Joint Guideline: Published 2016; Amended 2020
 (2) Market Research Conducted by Protara Therapeutics

NMIBC Represents the Most Common Form of Bladder Cancer

Bladder Cancer in the US

6th 
most prevalent
cancer in the U.S.⁽¹⁾

4x
more likely to be diagnosed
in **men**⁽²⁾



High rate of recurrence
with 3-year rate estimated at
up to **80%**⁽³⁾

NMIBC makes up
~80% of all bladder
cancer with **~65,000**
diagnosed per
year in the U.S.⁽⁴⁾

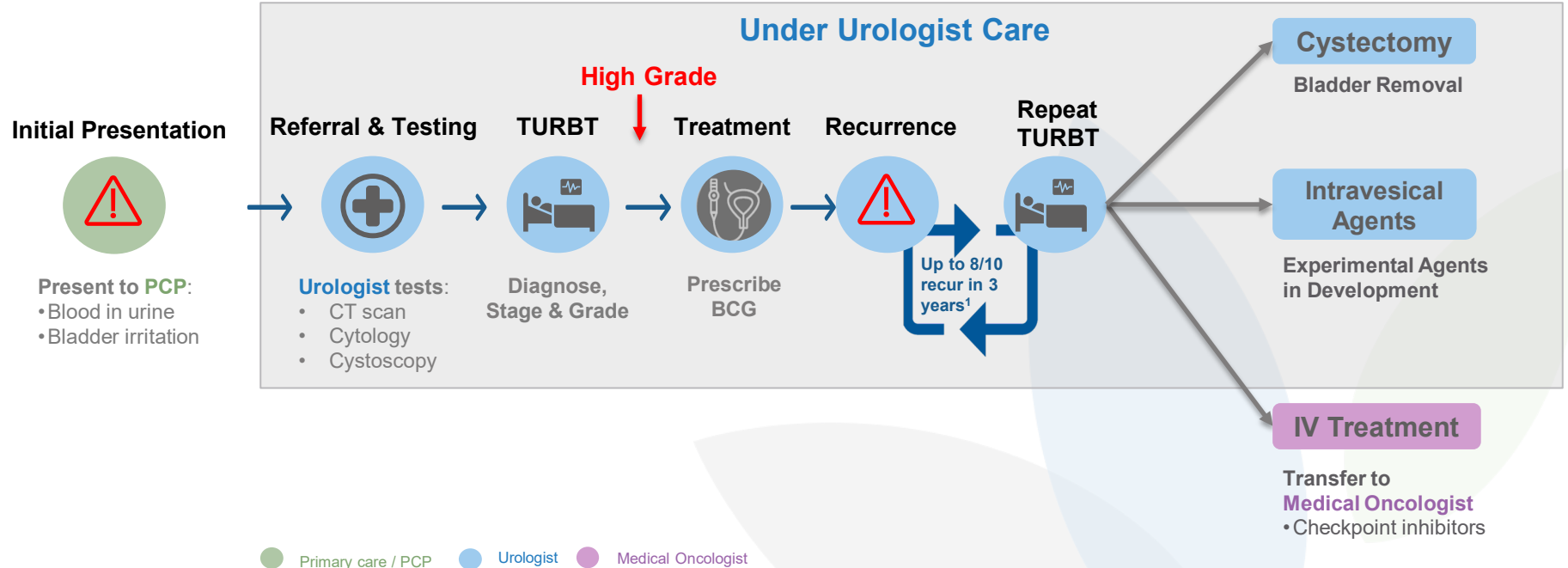
NMIBC patients
are treated by
a **urologist**



Significant increase in
recurrence, progression
& an escalated number
of patients needing
cystectomies⁽⁵⁾



Current Standard of Care Highlights High Unmet Need for Patients



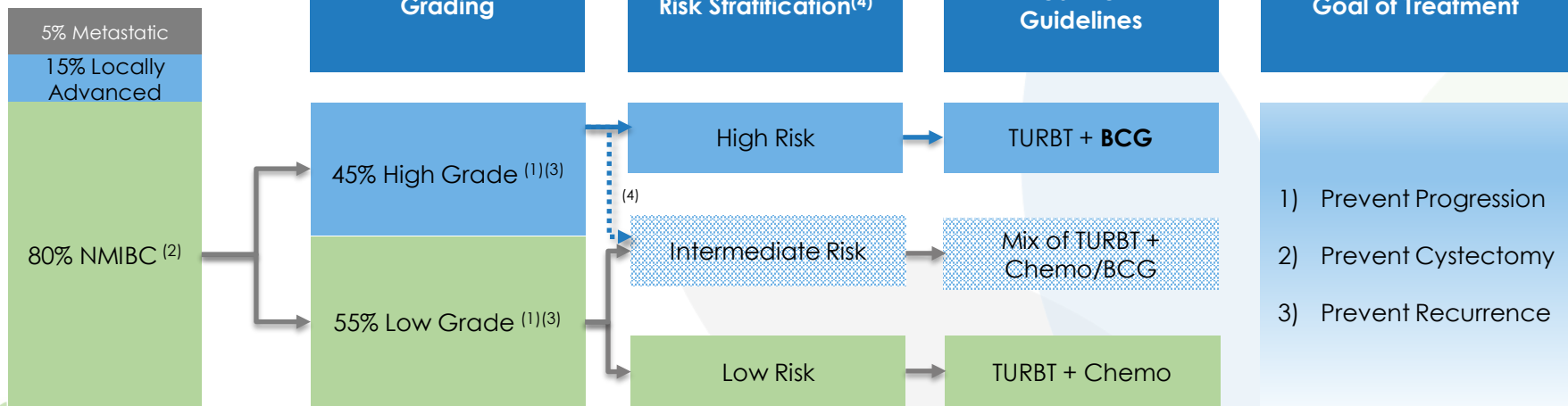
(1) Campbell Walsh 11th edition, Elsevier.

TARA-002 in NMIBC: Overview of U.S. NMIBC & Target Population

NMIBC is categorized and treated based on risk stratification, determined by combination of tumor grade, stage, size, recurrence history and focality

80K Patients Diagnosed with Bladder Cancer Annually⁽¹⁾
65K Patients with NMIBC⁽²⁾

Initial Protara targeted incident patient population:
~30K⁽¹⁾⁽²⁾⁽³⁾ Patients with High Grade NMIBC who are candidates for immunotherapy



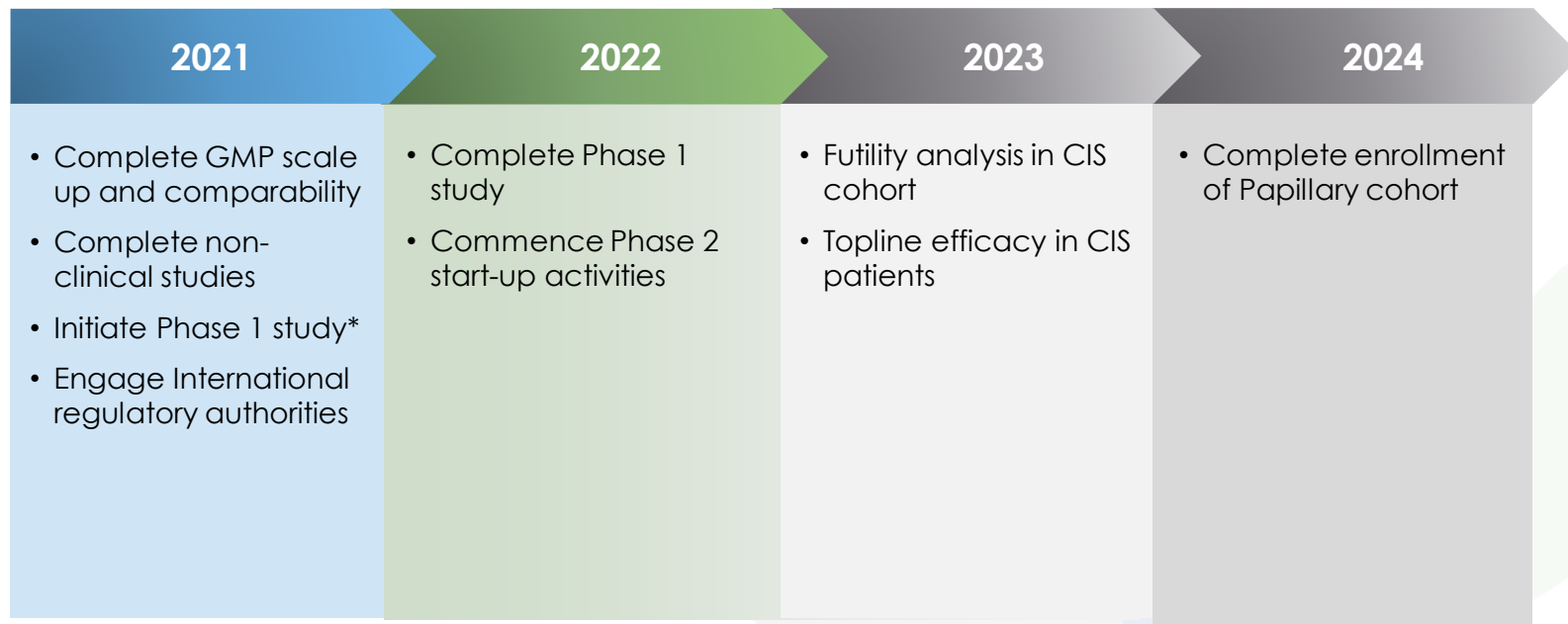
(1) National Cancer Institute, SEER Cancer Stat Facts: Bladder Cancer, 2019

(2) Anastasiadis et al. Therapeutic Advances in Urology, 2012

(3) Market Research Conducted by Protara Therapeutics

(4) American Urological Association risk stratification classifies some high-grade tumors as intermediate risk

TARA-002 in NMIBC: Estimated Development Timeline



TARA-002

LYMPHATIC MALFORMATIONS (LMs)



...d immunostimulatory cellular therapy
...nphatic Malformations.
Folate Met MAT1A MAT2A
CC[N+](C)(C)CCO.[Cl-]
THF
MTHFR
CHOLINE
A KEY FACTOR
essential nutrient
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of
Streptococcus pyogenes
line Chloride for IFALD - An
investigational phospholipid substrate
replacement therapy for intestinal
failure-associated liver disease (IFALD)

TARA-002 in LMs



Lymphatic Malformations

Rare, non-malignant lesions consisting of dilated, lymphatic fluid-filled sacs caused by abnormal development of the lymphatic endothelial system⁽¹⁾



Epidemiology

Epidemiology: incidence of lymphatic malformations is \approx 1,400-1,800 LM cases per year⁽²⁾



Current Treatment Options

Current treatment options include surgical excision with high complication (33%) and recurrence (55%) rates⁽³⁾ as well as off-label use of sclerosants



FDA Granted Pediatric Rare Disease Designation

Majority of LMs present at birth (65-75%) or by age 3 (80-90%) during active lymphatic growth period⁽³⁾



(1) Brouillard P, et al. J Clin Invest. 2014;124:898-904.

(2) Internal company estimates

(3) Ha J, et al. Curr Ped Rev. 2014;10:238-248.

OK-432 in LMs: Clear Evidence of Biologic Activity in Patients



Before

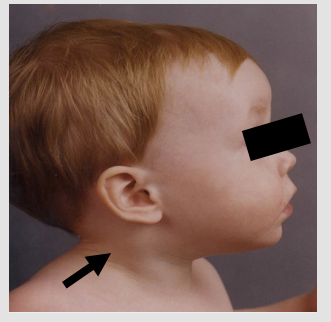
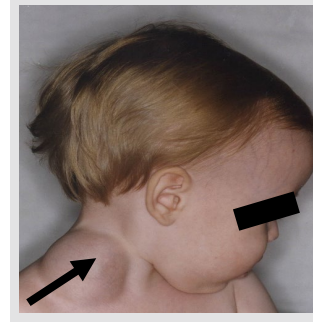
After



Before

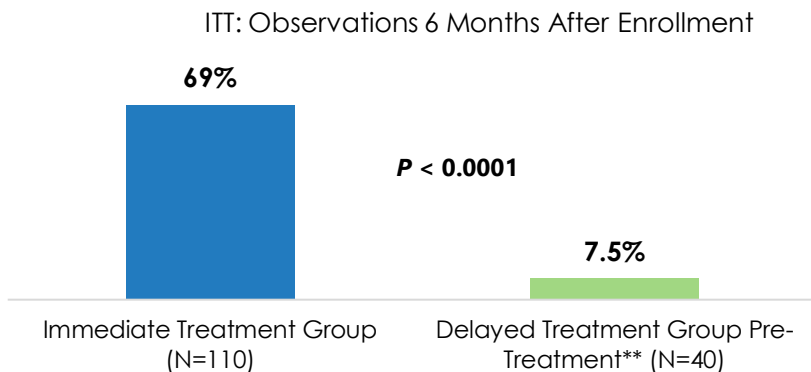


After

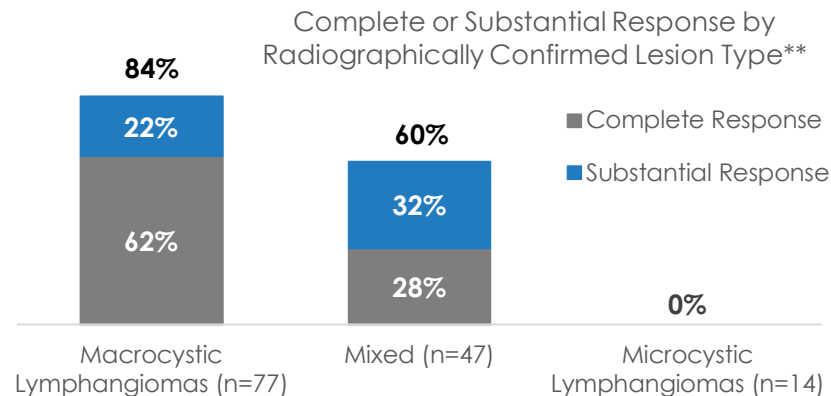


OK-432 in LMs: Robust Results of Completed Phase 2 Study⁽¹⁾ in U.S.

69% CLINICAL SUCCESS[‡] IN IMMEDIATE TREATMENT GROUP 6 MONTHS AFTER ENROLLMENT



84%* CLINICAL SUCCESS[‡] IN PATIENTS WITH MACROCYSTIC LESION TYPES



- During this same period, 7.5% of patients in the delayed treatment group experienced spontaneous regression of LM
- Treatment: 1-4 injections at 8-week intervals max of 0.2mg/session (2KE)

- Patients with radiographically confirmed macrocystic lesions had the greatest chance for clinical success
- In those patients with mixed lesions, clinical success was still achieved

[‡] Clinical Success was defined as complete or substantial response

*Reflects data prior to dosing with OK-432. After dosing, the clinical success rate was 66%, which was not statistically different from the Immediate Treatment Group

**Results were analyzed by lesion type across all treatment groups

(1) Results based on retrospective analysis of source verified data that included the full dataset of subjects enrolled in the P2 randomized study between January 1998 and August 2005, including data in the published study (Smith et al. 2009) which included subjects enrolled between January 1998 and November 2004.

OK-432 in LMs: Compelling Safety Record

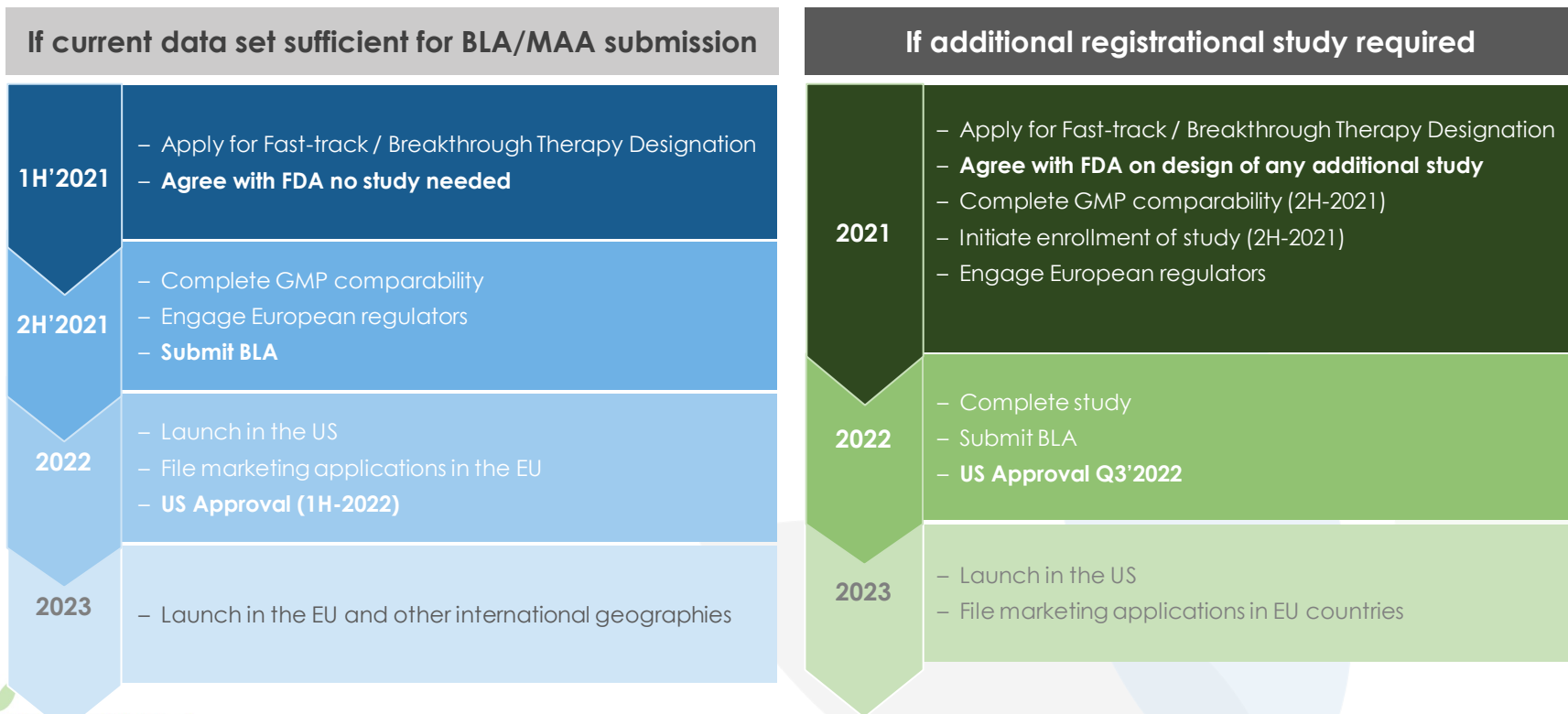
Safety Profile*

- Most common AEs with treatment were local injection site reactions, fever, fatigue, decreased appetite, with resolution within two weeks
- Treatment emergent SAEs **related** to OK-432: reported in 4.1% of patients, with the most severe events being airway obstruction and facial paralysis due to massive swelling post-injection that required tracheostomy and hospitalization. Both of these events were reported as resolved.
- One SAE **related** to OK-432 led to discontinuation: Proptosis of the eye
- One SAE **not related** to OK-432 led to death: Death due to tracheotomy tube obstruction

TARA-002 in LMs: Regulatory Update

- 1 IND update completed and filed with FDA Division of Vaccines and Related Products
- 2 Vaccines division began review of IND update in late Q4'2020 and requested a CSR summarizing the totality of the Iowa LMs Phase 2 study
- 3 CSR has been submitted and dialogue with Vaccines Division is ongoing
- 4 Company continues to prepare for the potential to file a BLA in 2H 2021 or to initiate additional clinical work in LMs as required

TARA-002 in LMs: Planned Next Steps

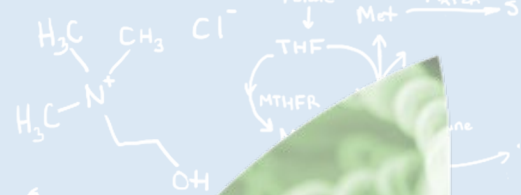


IV CHOLINE CHLORIDE

INTESTINAL FAILURE ASSOCIATED LIVER
DISEASE (IFALD)



...d immunostimulatory cellular therapy
...lymphatic Malformations.



CHOLINE
A KEY FACTOR
essential

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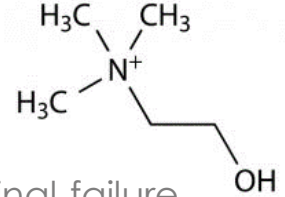
Choline Chloride for IFALD - An
investigational phospholipid substrate
replacement therapy for intestinal
failure-associated liver disease (IFALD)

IV Choline in IFALD: Late-stage Opportunity for an Unmet Medical Need



An Essential Molecule in Several Metabolic Processes

Patients dependent on Parenteral Nutrition (PN) cannot absorb sufficient levels of choline. Data confirms that choline deficient diets results in steatosis and cholestasis⁽¹⁾



Clinical History Supporting Choline Substrate Replacement in IFALD (intestinal failure associated liver disease) Patients

A Phase 2 study demonstrated the clinical potential of choline substrate replacement therapy by reversing certain hallmark pathologies of IFALD⁽¹⁾



Strong Market Opportunity with Potential to Expand Addressable Patients

Prevalence of patients on PN 79/million⁽²⁾; recent Medicare diagnosis data suggests ~ 5,000 IFALD patients⁽³⁾

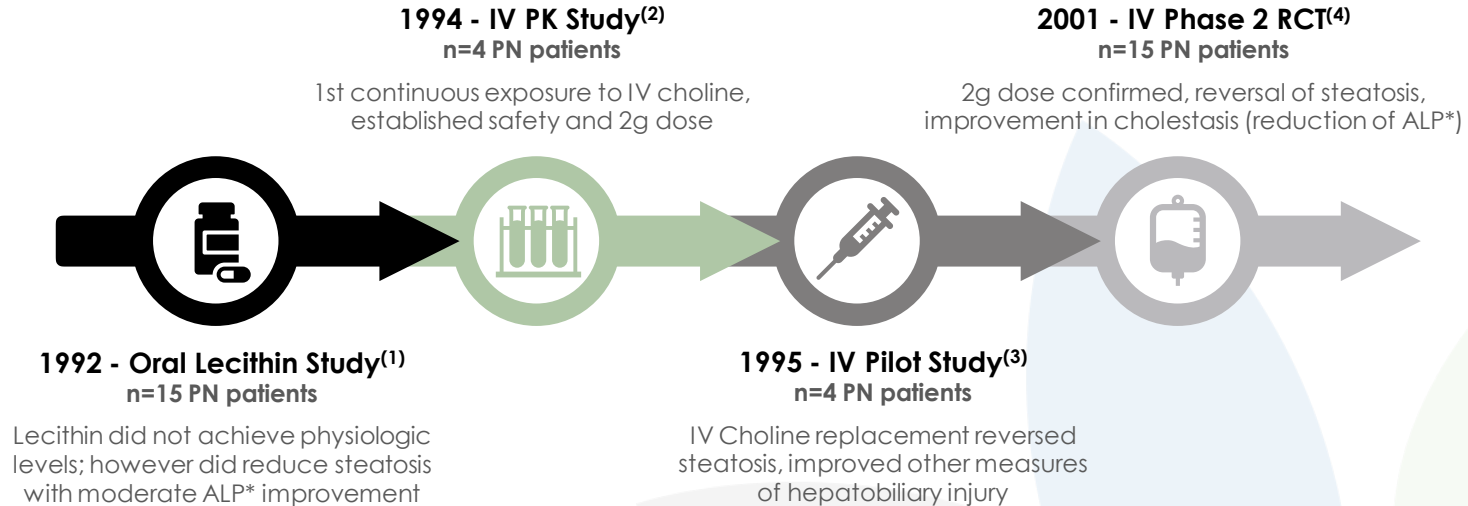


Clear Regulatory and Clinical Path Forward

FDA designations (Orphan Drug Designation, Fast Track Designation) combined with encouraging feedback from End of Phase 2 meeting for Phase 1 PK study followed by Phase 3 trial

IV Choline in IFALD: Informative Clinical History

A significant body of supportive evidence across 4 studies



IV Choline in IFALD: Multi-Center Phase 2a POC Study

Randomized, Controlled Study Design & Objective

IV CHOLINE REPLACEMENT PROOF OF CONCEPT STUDY ⁽¹⁾	
Study Design	Randomized Double-blind Phase 2 Trial
Subjects	15 (9 per protocol)
Age	>16 years old
PN Requirement	Greater than 80% of all nutrient requirements supplied by PN
Randomization	1:1 Usual PN or PN + 2g IV choline/Day
Duration of Treatment	24 Weeks
Visits	Weeks 2,4,6,12,16, 20, 24
Follow up	Week 34
Dose	2g Choline Chloride QD in PN solution

- The IV Choline Chloride replacement proof-of-concept, randomized study did not have pre-specified endpoints
- The primary objective of the study was to determine if IV Choline Chloride substrate replacement would reverse hepatic steatosis and improve liver function in patients receiving long-term parenteral nutrition (PN)

IV Choline in IFALD : Phase 2 Results

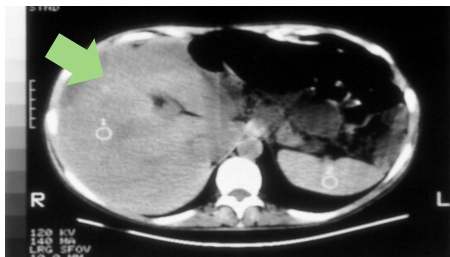
Improvement in Steatosis and Cholestasis

CLINICALLY MEANINGFUL IMPROVEMENT IN STEATOSIS

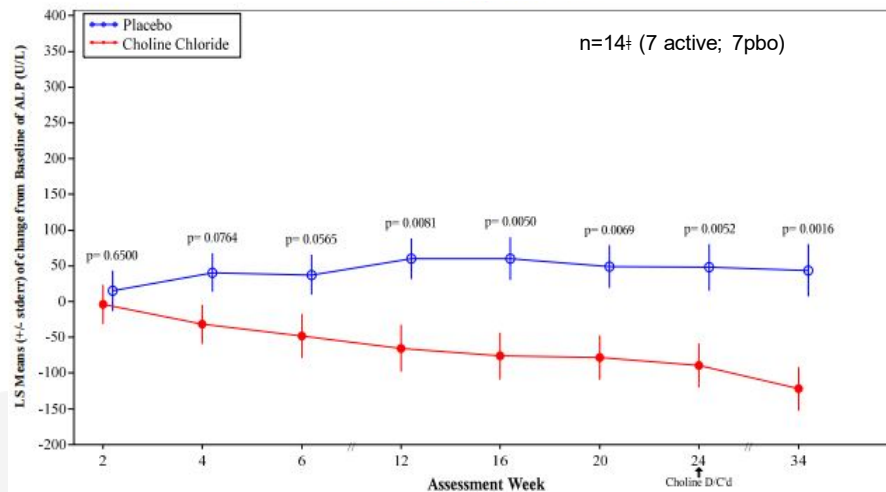
At Baseline



After 24 Weeks



CHOLESTASIS IMPROVEMENT: ALL PATIENTS*(1)



*Mixed model for repeated measurement (MMRM) method used

†A placebo subject was excluded from all analyses due to likely IV contrast-induced imaging abnormalities, confirmed by independent radiologist

IV Choline in IFALD: Prevalence Study

Prevalence study underway to enhance understanding of the patient population

DESIGN

Retrospective, observational study of patients in both academic & community settings

POPULATION

Patients dependent on PN for 6 or more months

OBJECTIVE

Understand presence/incidence of liver disease in this population to enhance value of development potential

QUESTIONS

How many individuals currently on service have been dependent on PN for 6 or more months?
What percentage of these have elevated ALP levels ($> 1.5 \times \text{ULN}$) as an indicator of liver disease?

Summary



Building Momentum in 2021

LMs:

Near-term Rare
Disease Opportunity

- Q1'21: Submitted CSR to FDA (Vaccines Division) to support TARA-002 for LMs with the potential to file our BLA in 2H 2021
- 2H'21: Complete GMP scale up and comparability

NMIBC:

Significant Market
Potential

- 1H'21: Complete nonclinical studies (tox, MOA, immunogenicity)
- 2H'21: File IND
- Late 2021: Initiate Phase 1 study*

IV Choline:

Late-Stage Pipeline
Opportunity

- 2H'21: Complete prevalence study to better characterize epidemiology of IFALD

Financials:

Estimated funding
through early 2023

- \$166M as of September 30, 2020
- 19.2M Common Share Equivalents: 11.2M Common + 8.0M Preferred on as-converted basis as of November 10, 2020

*Subject to acceptance of IND filing



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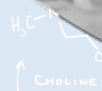
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