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Early- and Long-Term Outcome Data of Patients With Pseudomyxoma Peritonei From Appendiceal Origin Treated by a Strategy of Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy

Terence C. Chua, Brendan J. Moran, Paul H. Sugarbaker, Edward A. Levine, Olivier Glehen, François N. Gilly, Dario Baratti, Marcello Deraco, Dominique Elias, Armando Sardi, Winston Liauw, Tristan D. Yan, Pedro Barrios, Alberto Gómez Portilla, Ignace H.J.T. de Hingh, Wim P. Ceelen, Joerg O. Pelz, Pompiliu Piso, Santiago González-Moreno, Kurt Van Der Speeten, and David L. Morris

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Author affiliations appear at the end of this article.

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Corresponding author: Terence Chua, BScMed (Hons), MB, BS, Hepatobiliary and Surgical Oncology Unit, UNSW Department of Surgery, St George Hospital, Sydney, Australia; e-mail: terence.chua@unsw.edu.au.

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ABSTRACT

Purpose
Pseudomyxoma peritonei (PMP) originating from an appendiceal mucinous neoplasm remains a biologically heterogeneous disease. The purpose of our study was to evaluate outcome and long-term survival after cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) consolidated through an international registry study.

Patients and Methods
A retrospective multi-institutional registry was established through collaborative efforts of participating units affiliated with the Peritoneal Surface Oncology Group International.

Results
Two thousand two hundred ninety-eight patients from 16 specialized units underwent CRS for PMP. Treatment-related mortality was 2% and major operative complications occurred in 24% of patients. The median survival rate was 196 months (16.3 years) and the median progression-free survival rate was 98 months (8.2 years), with 10- and 15-year survival rates of 63% and 59%, respectively. Multivariate analysis identified prior chemotherapy treatment ($P < .001$), peritoneal mucinous carcinomatosis (PMCA) histopathologic subtype ($P < .001$), major postoperative complications ($P = .008$), high peritoneal cancer index ($P = .013$), debulking surgery (completeness of cytoreduction [CCR], 2 or 3; $P < .001$), and not using HIPEC ($P = .030$) as independent predictors for a poorer progression-free survival. Older age ($P = .006$), major postoperative complications ($P < .001$), debulking surgery (CCR 2 or 3; $P < .001$), prior chemotherapy treatment ($P = .001$), and PMCA histopathologic subtype ($P < .001$) were independent predictors of a poorer overall survival.

Conclusion
The combined modality strategy for PMP may be performed safely with acceptable morbidity and mortality in a specialized unit setting with 63% of patients surviving beyond 10 years. Minimizing nondefinitive operative and systemic chemotherapy treatments before definitive cytoreduction may facilitate the feasibility and improve the outcome of this therapy to achieve long-term survival. Optimal cytoreduction achieves the best outcomes.

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INTRODUCTION

Epithelial appendiceal neoplasms account for 1% of colorectal cancer.[1,2] In its early stages, the diagnosis may be made incidentally at the time of appendectomy, occurring in less than 1% of appendectomies.[3] Advanced disease is often a result of tumor perforation and seeding of tumor cells within the peritoneal cavity leading to the clinical syndrome of pseudomyxoma peritonei (PMP). In the past, management of this disease involved repeated drainage of mucinous ascites or surgical debulking through removal of the primary tumor and omental mass. In an article from the Mayo clinic, Gough et al[4] reported that 34% of patients with limited low-grade appendiceal pseudomyxoma could become free of disease via debulking surgery, with an estimated 10-year survival rate of 32%. In another article from the