The Natural History of Retroperitoneal-Abdominal-Pelvic Ganglioneuromas: An International Study By The Transatlantic Australasian Retroperitoneal Sarcoma Working Group (TARPSWG)

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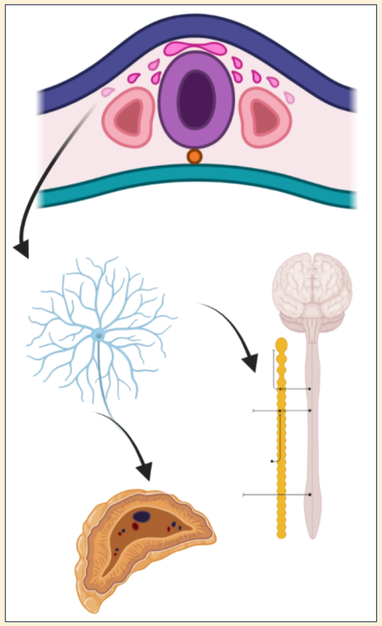
No conflicts of interest to declare.

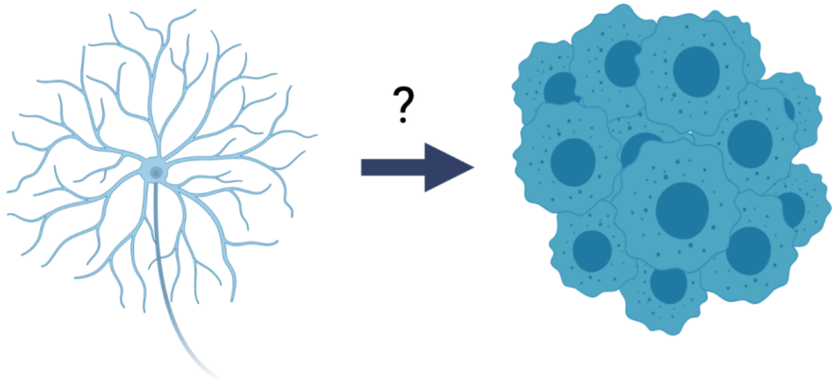
INTRODUCTION

- Ganglioneuromas (GN) are rare tumors derived from neural crest cells within the sympathetic plexus and adrenal glands
- Often discovered incidentally, or during work-up of non-specific symptoms caused by mass effect
- Occasionally occur in setting of hereditary syndromes (NF-1, MEN2)



INTRODUCTION

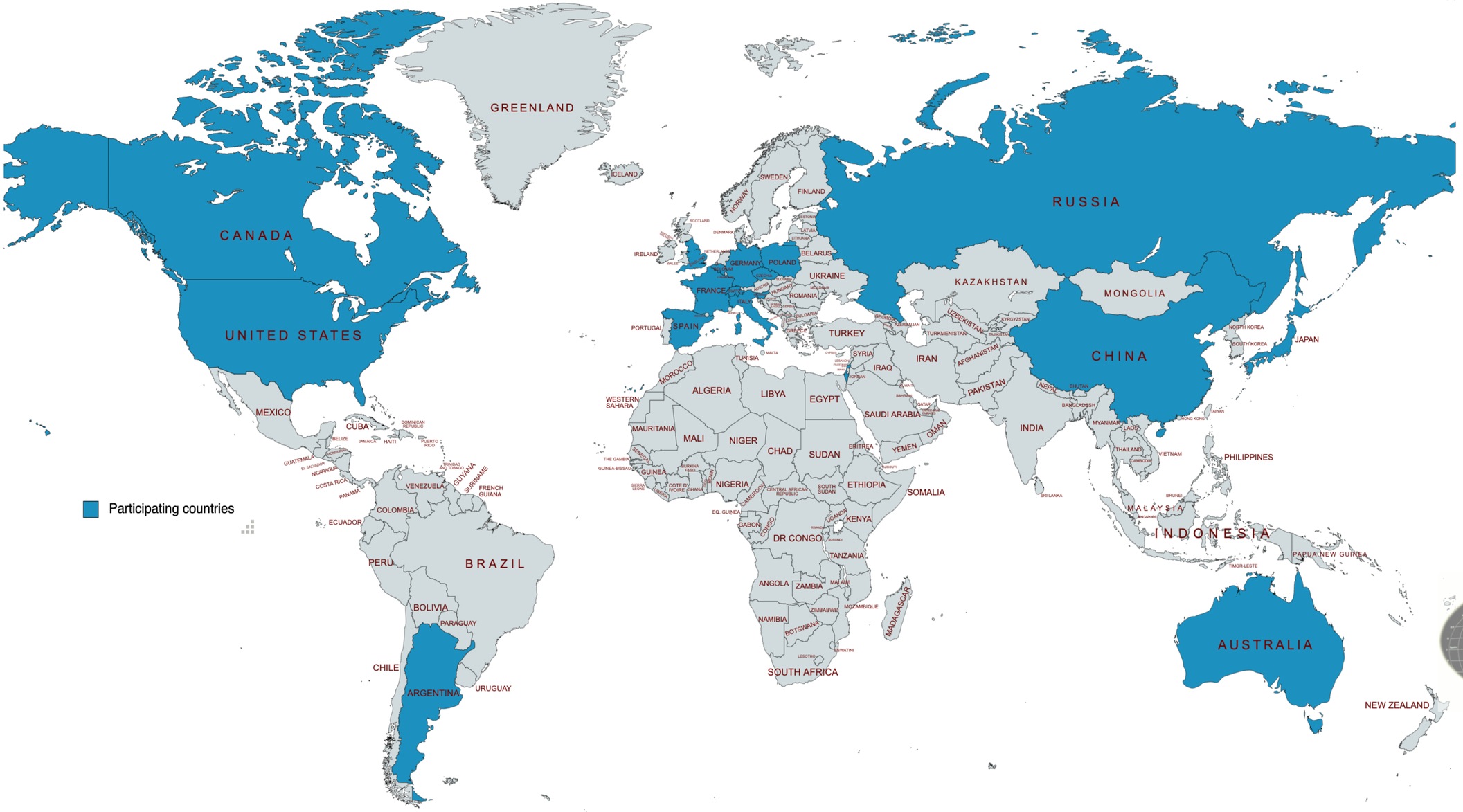
- Data on Retroperitoneal-Abdominal-Pelvic GN is limited to case reports and single-institution case series
- GN lack specific clinical symptoms/signs, laboratory findings, or diagnostic imaging characteristics
- Rates of malignant degeneration to neuroblastoma remain unclear



OBJECTIVE

- To conduct a retrospective, multi-institutional, multi-national study of the natural history and clinical management of ganglioneuroma.

GLOBAL COLLABORATION:
29 INSTITUTIONS ACROSS 5 CONTINENTS



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METHODS

- Following IRB approval, retrospective chart reviews of patient diagnosed with retroperitoneal, abdominal, and pelvic GN were performed
- January 2000 – January 2020
- All ages included
- Excluded head and neck, mediastinal, and GI tract tumors

- Examined patient demographics, clinicopathological features, imaging follow up, approaches to operative & non-operative management, and patient outcomes

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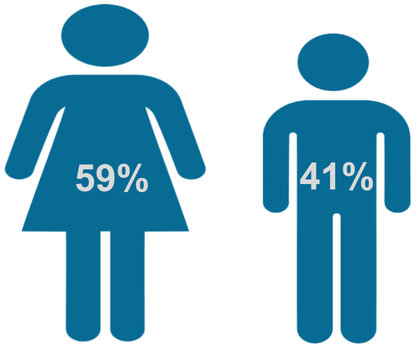
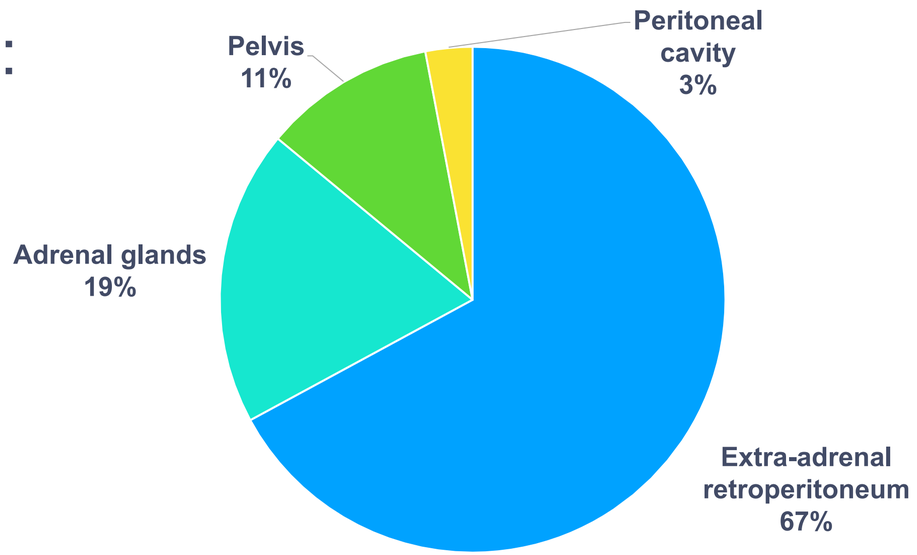
PATIENT DEMOGRAPHICS

- 328 patients from 29 institutions (5 continents)
- Median age at diagnosis: 37 yo (range, . – 79 yo)
- Sex: 59% female, 41% male
- Location:





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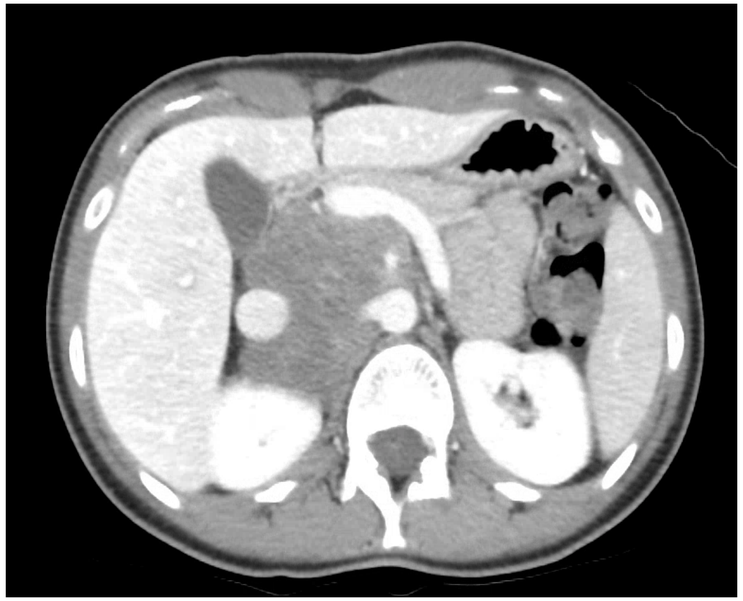
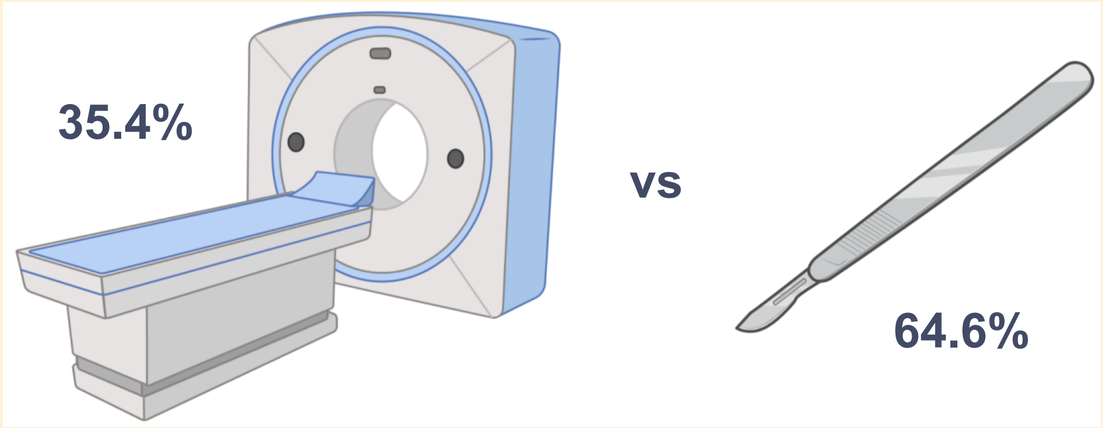
CLINICAL FEATURES

- Symptomatic presentation: 41%
- Vascular encasement: 17%
- Major nerve involvement: 8%
- Hereditary syndromes: 2.4%
    - Neurofibromatosis-1 (N=6)
    - MEN 2A (N=1)
    - Schwannomatosis (N=1)



MANAGEMENT: NON-OPERATIVE VERSUS OPERATIVE

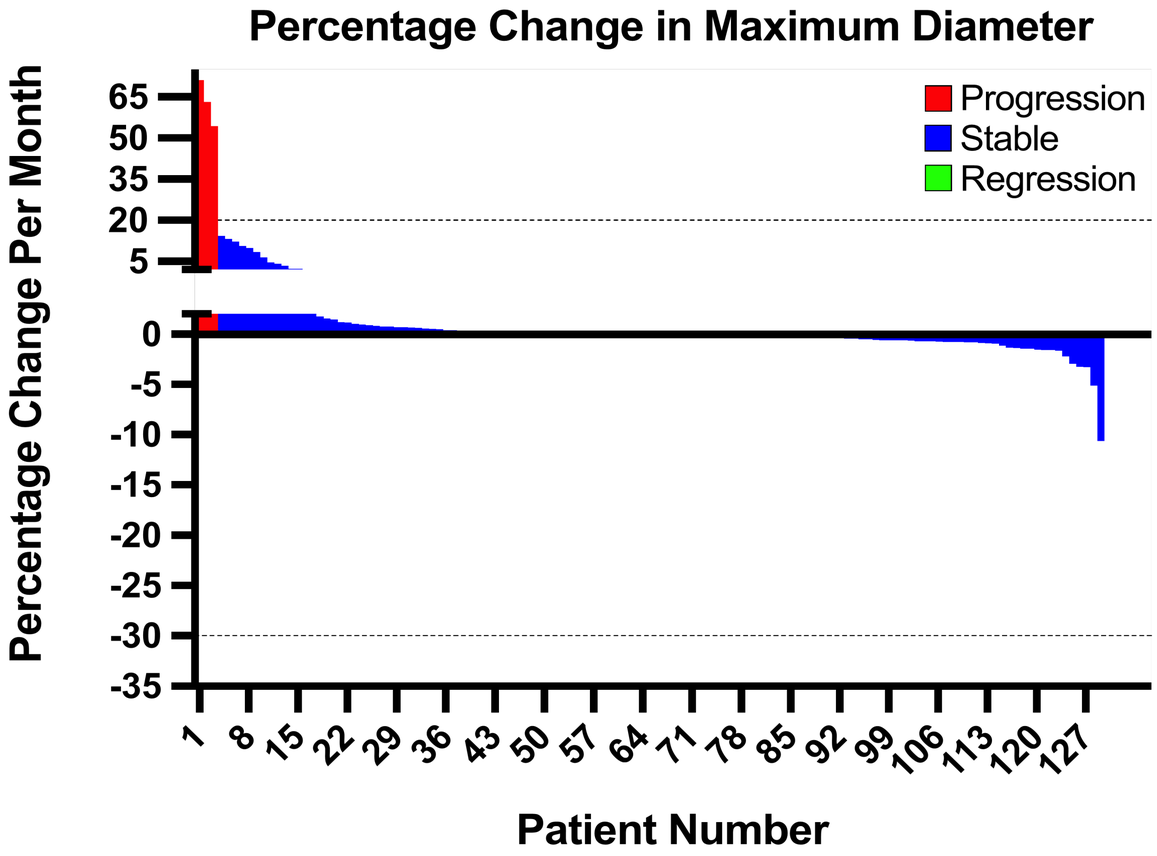


- Reasons for non-operative management:
  - Asymptomatic
  - Indolent nature of the lesion
  - Potential risks of resection

- Median follow-up of 1.9 years (IQR 0.8 – 4.2)

TUMOR SIZE AND SURVEILLANCE

Median Tumor Diameter	7.2 cm (IQR 5.0 – 9.9)
Vascular encasement	9.4 cm (IQR 7.2 – 12.0)



Progressive disease in 3 (2.3%)

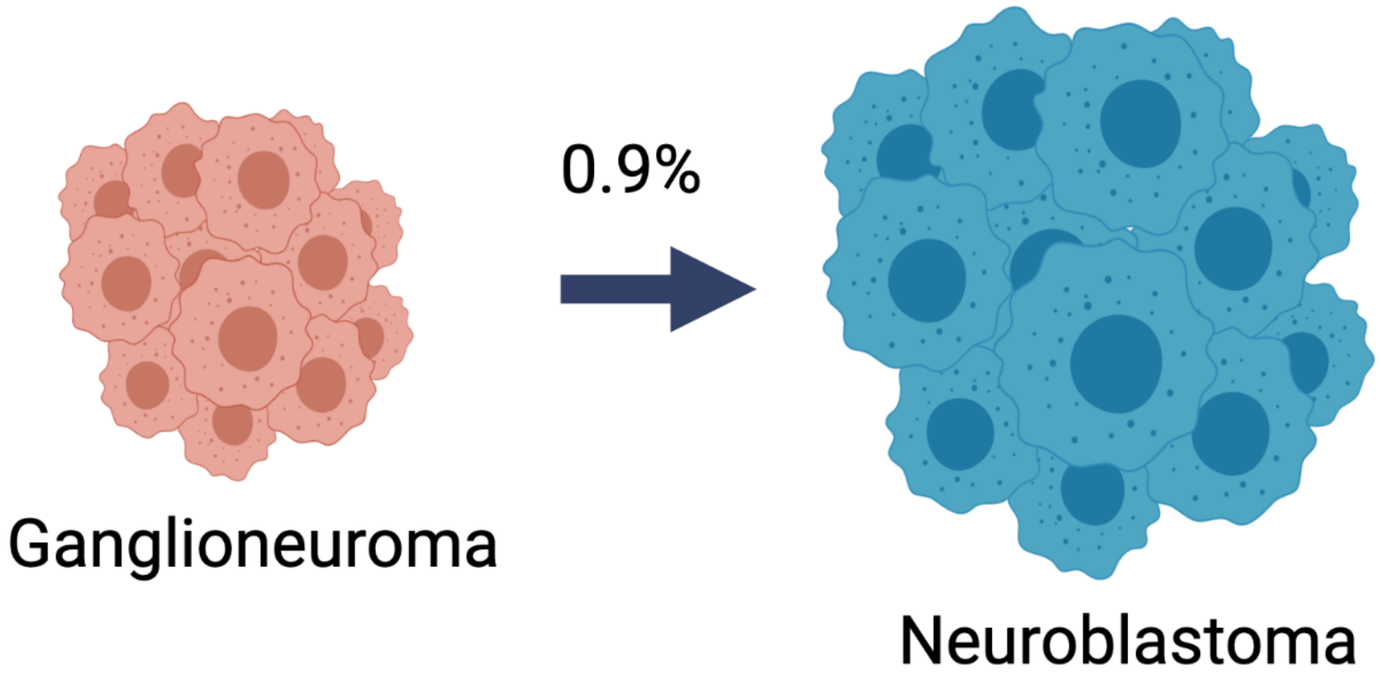
Stable disease in 126 (97.7%)

OPERATIVE MANAGEMENT

- Indications: Tumor growth, symptoms/mass effect, concerns for malignancy, and diagnostic uncertainty
- 64.6% underwent resection
    - 74.5% achieving R0/R1 resection
- 84% disease free at median follow-up of 3.0 years (IQR 1.0 – 5.5)
- Recurrences: 5 (2.4%) cases

NEUROBLASTOMA TRANSFORMATION

Neuroblastoma transformation	3 (0.9%)
Neuroblastoma transformation age	9-11 yo



SUMMARY

- Most patients with ganglioneuroma present sporadically
- Less than half of patients present with symptoms
- Most GN have indolent disease courses and <1% transformed into neuroblastoma
- Non-operative management with serial imaging evaluations may be appropriate when the risks outweigh benefits of resection in confirmed benign or asymptomatic cases
- If technically feasible and safe, definitive operative management with R0/R1 resection may be recommended when tumors are symptomatic, growing or of uncertain biology

CONCLUSION

- Largest study of ganglioneuroma to date
- While retrospective in nature, we can utilize this data to learn about the natural history of this tumor type to help guide management decisions and to design future studies

TARPSWG COLLABORATIVE

Carolyn Nessim, Laura Nguyen, Emily Z. Keung, Christina L. Roland, Dirk Strauss, Gausihi Sivarajah, Marco Fiore, Davide Biasoni, Stefano Cioffi, Ferdinando Carlo Maria Cananzi, Federico Sicoli, Vittorio Quagliuolo, Jun Chen, Chenghua Luo, Rebecca Gladdy, Carol Swallow, Wendy Johnston, Samuel J. Ford, Caroline Evenden, Fabio Tirotta, Max Almond, Piotr Rutkowski, Marika Krotewicz, Elisabetta Pennacchioli, Kenneth Cardona, Adriana Gamboa, Daphne Hompes, Attila Kollár, Christoph O. Ryser, Nikolaos Vassos, Chandrajit P. Raut, Mark Fairweather, Dagmar Adamkova Krakorova, Sergio Damián Quildrian, Andraz Perhavec, Eran Nizri, Jeffrey M. Farma, Stephanie H. Greco, Bruno Vincenzi, José Antonio González Lopez, Mireia Solans Solerdecoll, Shintaro Iwata, Suguru Fukushima, Teresa Kim, Francesco Tolomeo, Hayden Snow, Ynez Howlett-Jansen, Dimitri Tzanis, Maxim Nikulin, Alessandro Gronchi

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Humanitas University, Humanitas Clinical and Research Center/ Italy
IEO European Institute of Oncology/ Italy
Inselspital, Universitätsspital Bern/ Switzerland
Institut Curie/ France
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