



Dipartimento Rete Oncologia del Piemonte  
e della Valle d'Aosta



*Incontro del GIC*  
***Tumori rari***  
***G.I.S.T. e N.E.T.***

**IL SOSPETTO DI N.E.T. E LE INIZIATIVE PER RENDERE PIU'  
TEMPESTIVA LA DIAGNOSI**

**Dr.ssa Nadia Birocco**  
**SC Oncologia 1**  
**COES**



# IL SOSPETTO DI N.E.T. E LE INIZIATIVE PER RENDERE PIU' TEMPESTIVA LA DIAGNOSI



- *Fonti bibliografiche*
- *Epidemiologia*
- *Perchè il sospetto*
- *Perchè diagnosi tempestiva*
- *Le iniziative*



# DAL GENERALE.....



## Long-term Survival of Patients with Small Intestinal Carcinoid Tumors

Niklas Zar, M.D.,<sup>1,3</sup> Hans Garmo, Ph.D.,<sup>2</sup> Lars Holmberg, M.D., Ph.D.,<sup>2</sup> Jonas Rastad, M.D., Ph.D.,<sup>3</sup>  
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American Journal of Gastroenterology  
© 2007 by Am. Coll. of Gastroenterology  
Published by Blackwell Publishing

ISSN 0002-9270  
doi:10.1111/j.1572-0241.2007.01185.x

## A Three-Decade Analysis of 3,911 Small Intestinal Neuroendocrine Tumors: The Rapid Pace of No Progress

Irvin M. Modlin, M.D., Ph.D., D.Sc., Manish C. Chatterjee, B.S., Anthony K.C. Chan, B.Sc.,  
and Mark Kidd, Ph.D.

*Gastrointestinal Pathobiology Research Group, Yale University School of Medicine, New Haven, Connecticut*

## One Hundred Years After "Carcinoid": Epidemiology of and Prognostic Factors for Neuroendocrine Tumors in 35,825 Cases in the United States

James C. Yao, Mural Hassan, Alexandria Phan, Cecile Dagohoy, Colleen Lavery, Jannette E. Marrs,  
Eddie K. Abulula, Jason B. Fleming, Juan-Nicolas Vassihay, Asif Rashid, and Douglas B. Evans

VOLUME 26 - NUMBER 16 - JUNE 20 2008

JOURNAL OF CLINICAL ONCOLOGY

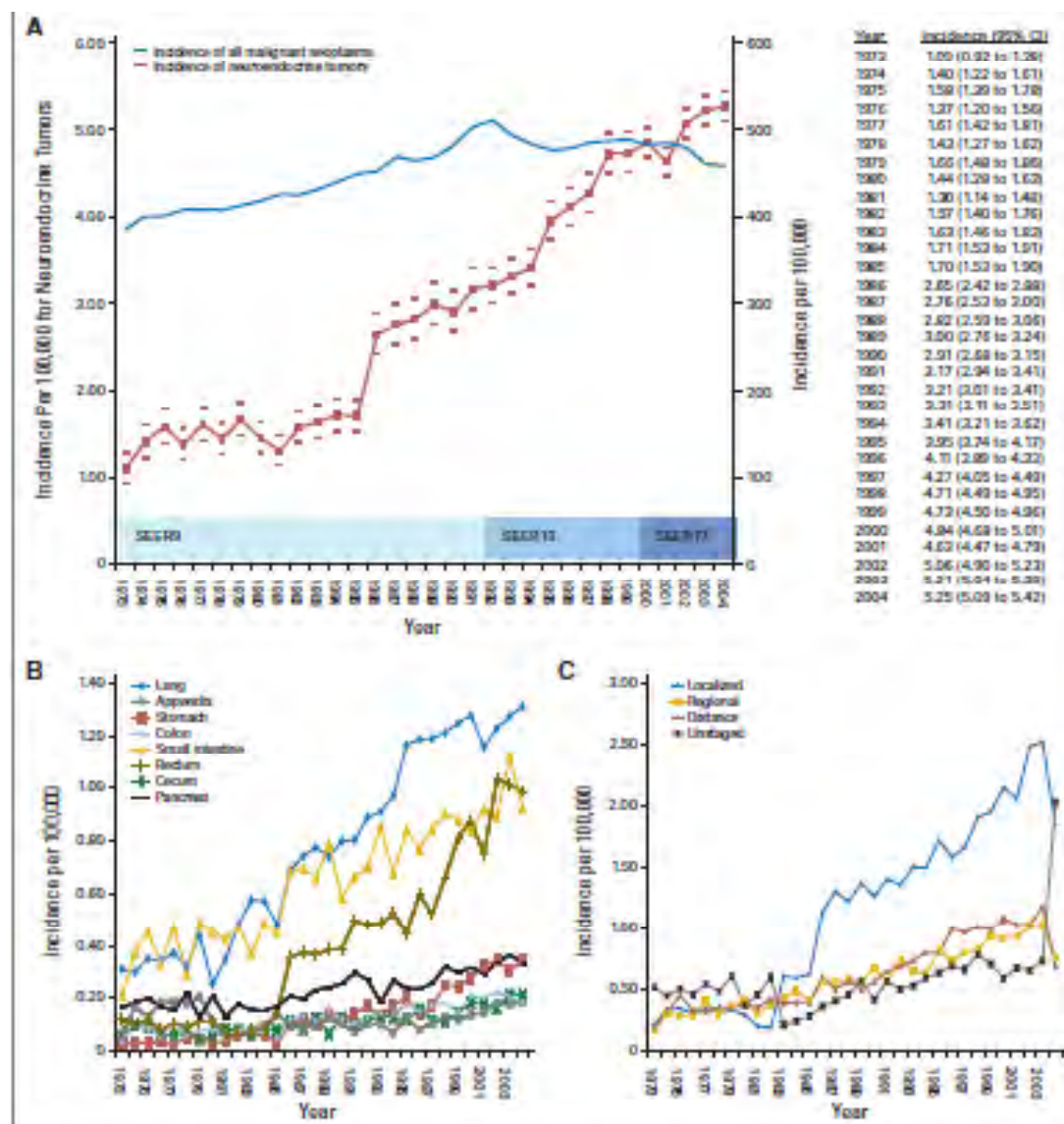
REVIEW ARTICLE

## EDITORIALS

## Measuring a doctor's performance: personality, health and well-being

*Occupational Medicine* 2006;56:438-441  
doi:10.1093/occmed/kqj076

# INCIDENZA



**Fig 1.** These graphs show the incidence of neuroendocrine tumors (NETs) over time, by site and by disease stage. (A) Annual age-adjusted incidence of NETs by year (1973 to 2004). The incidence is presented as the number of tumors per 100,000 (with 95% CI) age-adjusted for the 2000 US standard population. Cases were selected from the Surveillance, Epidemiology, and End Results database (1973 to 2004) using International Classification of Diseases for Oncology histology codes 8150 to 8157, 8240 to 8245, and 8260. (B) Time-trend analysis of the incidence of NETs by primary tumor site (1973 to 2004). Statistically significant increases in incidence at all sites are shown ( $P < .001$ ). (C) The incidence of NETs by disease stage at diagnosis. Statistically significant increases in incidence at all stages are shown ( $P < .001$ ).

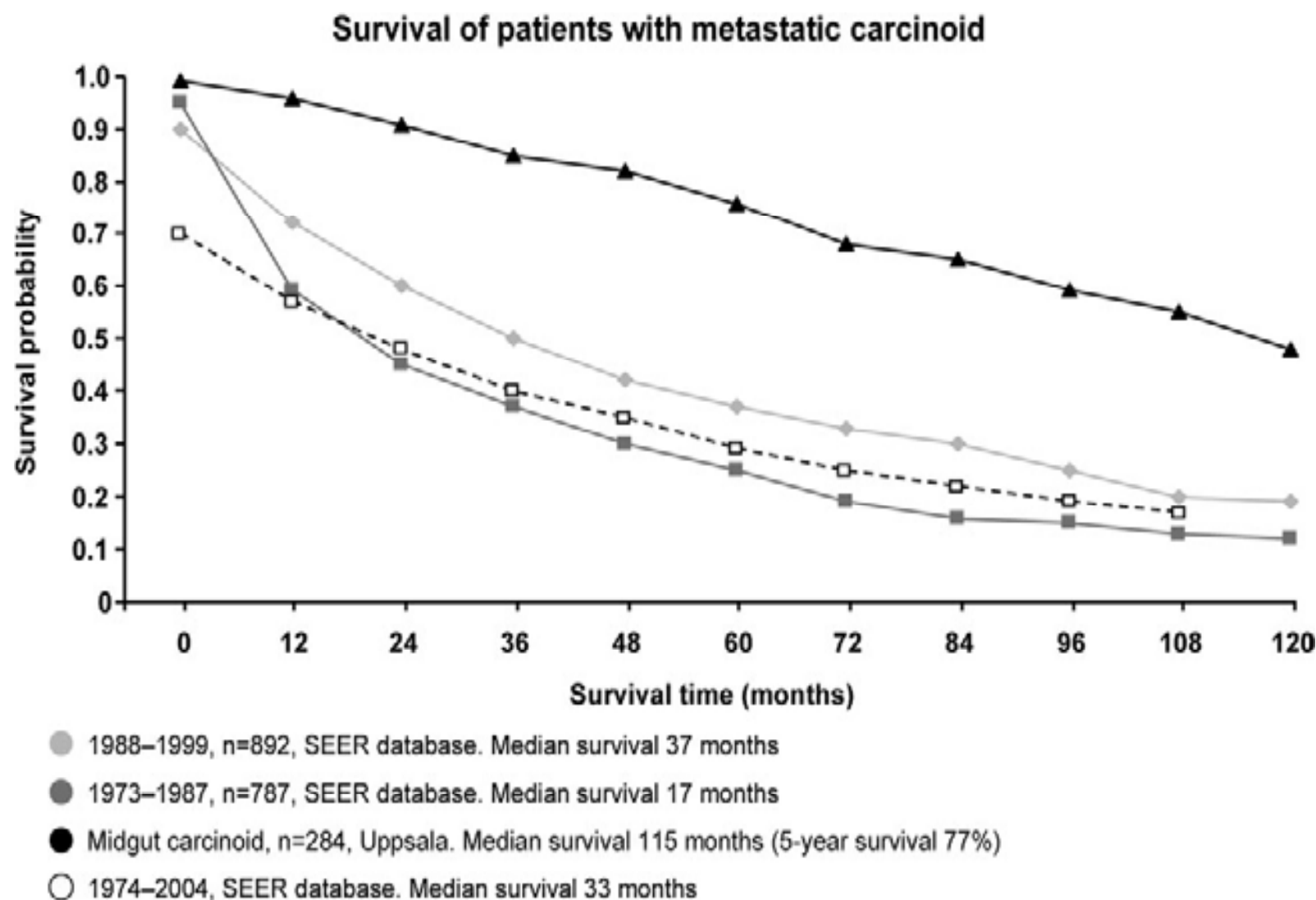


**INCIDENZA 5,25/100000**

**PREVALENZA 35/100000**

# I TUMORI NEUROENDOCRINI

K.E.Oberg, Annals of Oncology 21 (Supplement 7): vii72–vii80, 2010





# IL SOSPETTO



### Ipergastrinemia: diagnosi differenziale

gastrinoma	++++	(> 1000 pg / ml)
gastrite atrofica	++++	
iperplasia G antrale	++++	(> 150 pg / ml)
anemia pernicioasă	++++	

**Gastroenterologo,  
Cardiologo,  
Ginecologo,  
Dermatologo,  
Endocrinologo,  
Neurologo,  
Pneumologo  
Psicologo.....**

## DIARREA CRONICA REFRATTARIA A DIAGNOSI DIFFERENZIALE

- INSUFFICIENZA PANCREATICA
  - EPATOPATIA CRONICA
  - SCLEROSI SISTEMICA
  - DIABETE MELLITO
  - COLON IRRITABILE
  - INFEZIONI INTESTINALI (Batteri)
  - MORBO DI CROHN
  - MORBO CELIACO
  - JATROGENA
- (Reazioni Iatrogeniche: Enterici/Pan-Enterici, Lussuativi, Bismutici, Citostatici)
- AIDS
  - TUMORE NEUROENDOCRINO GEP



## Al particolare.....



### IL SOSPETTO MANCATO!

- Donna 44 anni, diarrea acquosa, astenia, vomito
- Durata sintomi: 8 mesi
- VIP sierico alla diagnosi: 119 ng/ml
- Carcinoma neuroendocrino ben differenziato del pancreas pT3N1 ki 67: 1%;

----->mts epatiche a 2 anni dalla diagnosi





## Al particolare.....



### IL SOSPETTO MANCATO!

- Donna 68 anni, dolori addominali ricorrenti, episodi di nausea , vomito e diarrea
- Durata sintomi: circa 1 anno
- Accesso in PS per occlusione intestinale (diagnosi precoce!)
- Carcinoma neuroendocrino intestino tenue, ki 67:2%,pT3N1; diffusa embolizzazione vascolare e perineurale
- ----->mts epatiche a 1 anno da diagnosi



# Al particolare.....



## IL SOSPETTO STUDIATO:

Donna 27 anni

Dolore addominale; effettua subito ECO ADD. SUP.

Pancreas con piccola formazione ipoecogena di 1 cm,  
lesione renale 2 cm

**Tumore neuroendocrino del pancreas pT1N0,  
neoplasia a cellule chiare del rene pT1N0**

-----> **sindrome VHL**



# **COSA IMPARARE**



**Esami di I livello (ecografia)**

**Endoscopia**

**Riconoscimento forme familiari**

**---->screening**

# DIAGNOSI TEMPESTIVA?

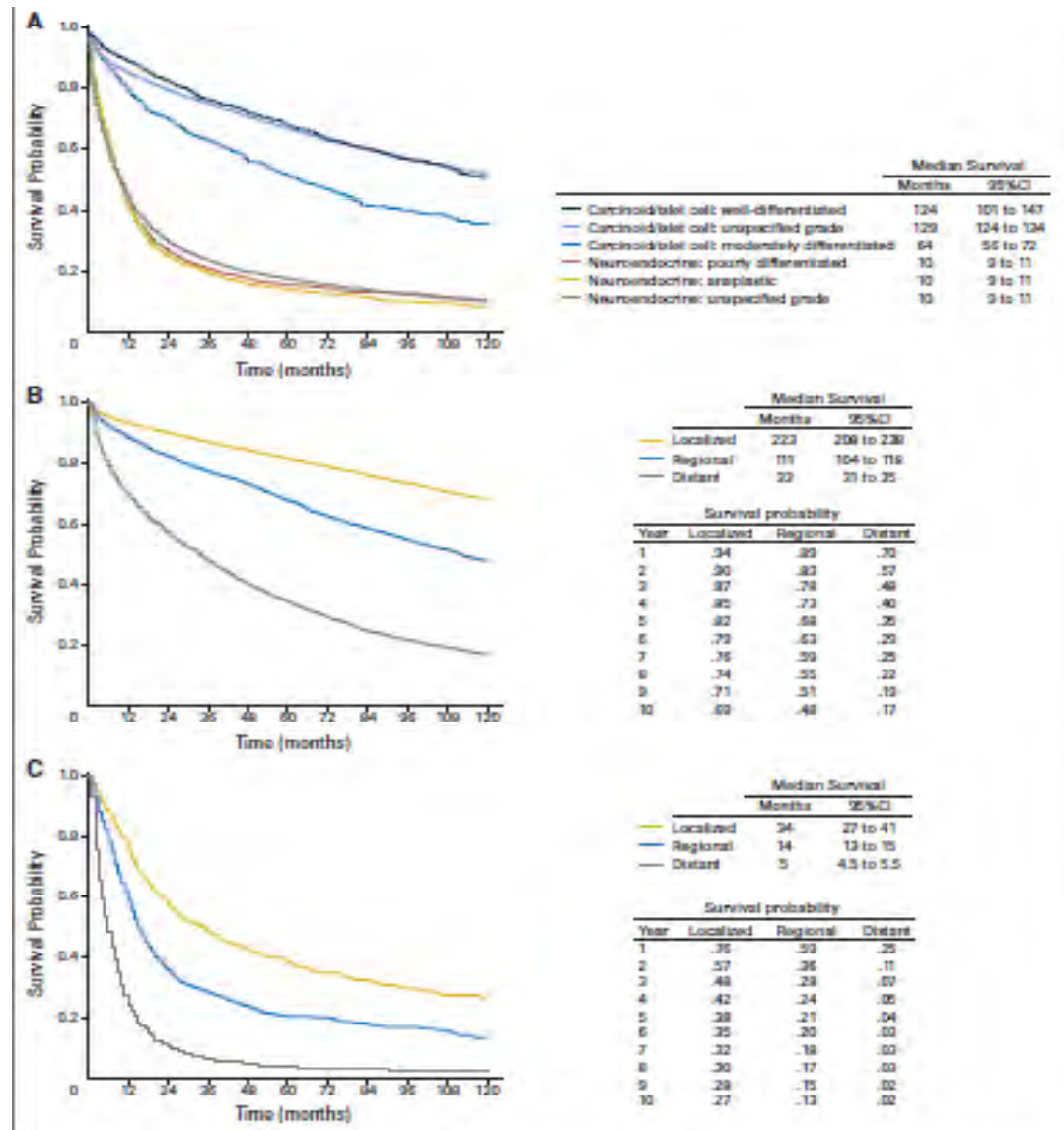
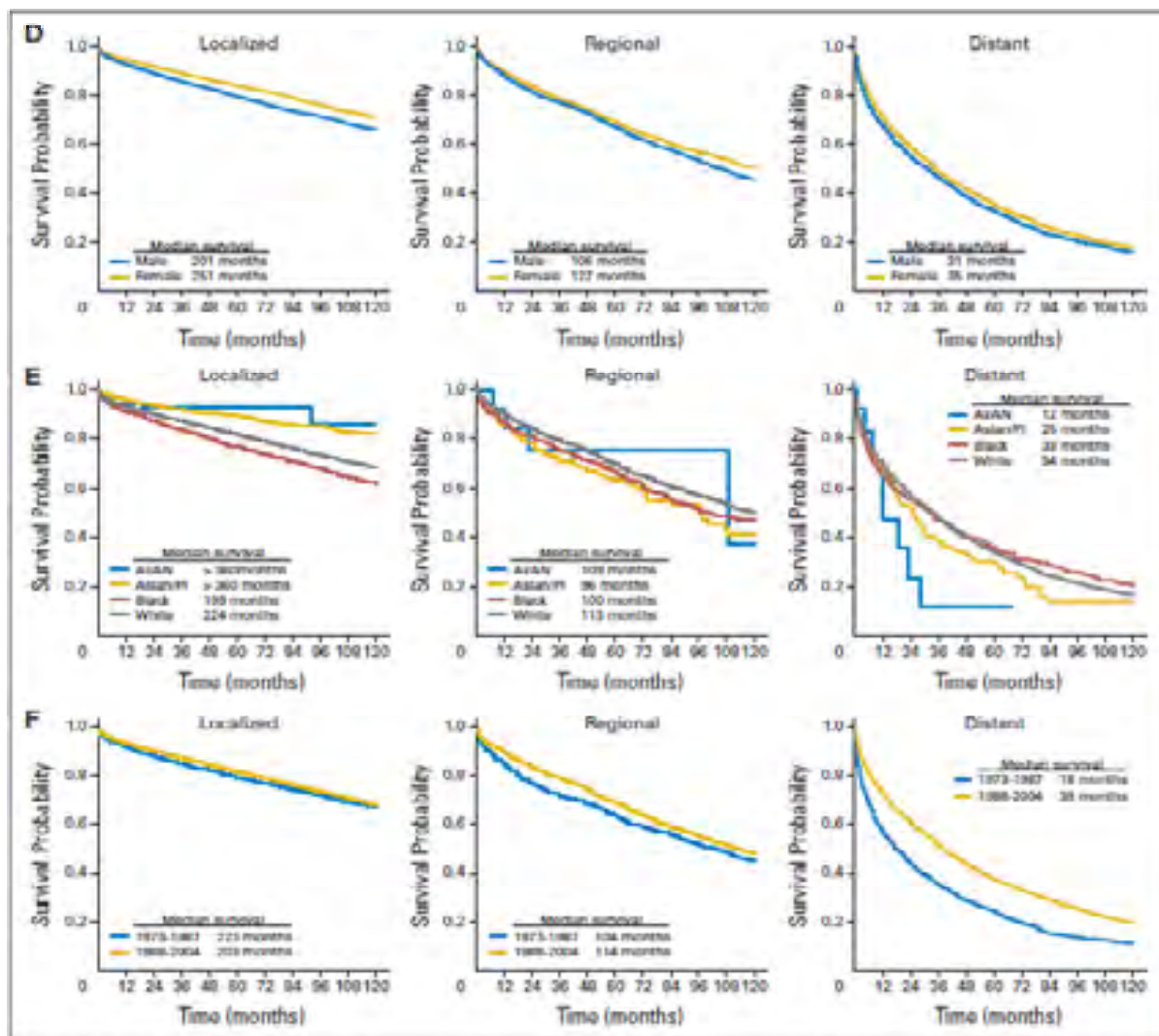


Fig 2. Survival duration by (A) histology (B) well- and moderately differentiated histology, and (C) poorly differentiated histology. Neuroendocrine tumor cases identified at autopsy or solely on the basis of death certificate were excluded. Median survival durations are presented in months (with 95% CI).

# DIAGNOSI TEMPESTIVA



**Fig 3 (Continued).** (C) Survival duration by age at diagnosis. Patients were separated into three groups according to their age at diagnosis (< 20, 21 to 60, and > 60 years). Age was found to be a strong predictor of outcome ( $P < .001$ ). (D) Survival duration by sex. Women had statistically significantly longer survival durations over all three categories histologies ( $P < .001$ ). (E) Survival duration by race. Patients were separated into four categories on the basis of race (American Indian/Alaskan Native (AIAN), Asian/Pacific Islander (Asian/Pi), African American, and white). American Indian/Alaskan Native and Asian/Pacific Islander patients had the longest survival durations for localized tumors, whereas white patients had the longest survival durations for metastatic diseases. (F) Survival duration by period of diagnosis. Patients were separated into two groups by year of diagnosis (1975 to 1987 and 1988 to 2004). Patients with metastatic disease had an improvement in median survival duration ( $P < .001$ ; from 18 to 38 months). There were no significant improvements in survival duration among patients with localized or regional disease. Each set of three graphs shows localized, regional, and distant survival from left to right.

**DIAGNOSI TEMPESTIVA**

**=**

**CURA TEMPESTIVA**



# One Hundred Years After “Carcinoid”: Epidemiology of and Prognostic Factors for Neuroendocrine Tumors in 35,825 Cases in the United States

*James C. Yao, Manal Hassan, Alexandria Phan, Cecile Dagothy, Colleen Leary, Jeannette E. Mares, Eddie K. Abdalla, Jason B. Fleming, Jean-Nicolas Vauthey, Asif Rashid, and Douglas B. Evans*

## Conclusion:

***Clinicians* need to become familiar with the natural history and patterns of disease progression, which are characteristic of these tumors.**



I

INIZIATIVE



# DAL PARTICOLARE....

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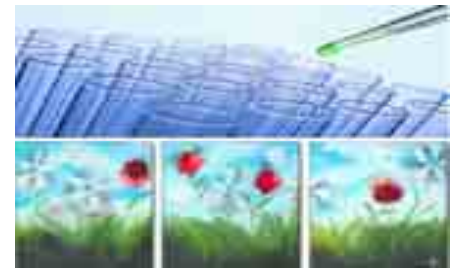
*Incontro del GIC*  
**Tumori rari**  
**G.I.S.T. e N.E.T.**



- **Gli strumenti:**
  - piattaforma virtuale dinamica, concreta, aperta
  - PDTA semplificati, “semi-automatici”
- **Gli operatori:**
  - gruppi di lavoro dedicati, coesi, anche sovra-aziendali
- **I numeri**



# AL GENERALE...



- Divisione dei compiti (ospedale, territorio)
  - Non sprecare informazioni

*“Under performance”= sprecare risorse*

Rispetto di limiti e “talenti” degli operatori  
(unicità di medico e paziente)



GRAZIE