

The Niigata Registry for Gut-Pancreatic Endocrinomas

Maintenance, Analysis and Supply with Windows Version

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Abstract

The purpose of the Niigata Registry for Gut-Pancreatic Endocrinomas (NRGPE) is to maintain information regarding gut-pancreatic endocrinomas and to make this information available to researchers engaged in the study of gut-pancreatic endocrinology. The Registry contains information that has been collected from sixty-five countries and regions worldwide in sixteen different languages. At present the Registry contains approximately 17,000 individually documented cases, 15,500 references and 28,000 reports, the latter including review articles and case studies that have been either duplicated or insufficiently well-documented. The specific requirements of individual researchers are served by the NUS System database, through which data can be computer-analyzed and measured against twenty-one basic tables for statistical analysis. Approximately 42% of all cases of endocrinocarcinoma are Japanese in origin and it is possible to determine the comparable features of both Japanese and foreign cases. The Niigata Registry, and its NUS System as well as a large quantity of the relevant literature, was donated to Niigata University School of Medicine and is housed in the Asahimachi Branch of the Niigata University Library. It is open to all researchers working in the field of gut-pancreatic endocrinology.

Key Words

Niigata registry, NUS system, endocrinocarcinoma, carcinoid, neuroendocrine carcinoma, endocrine cell carcinoma

Introduction

Several other Registries, in addition to the Niigata Registry store data that is used in carcinoid analysis. Located mainly in the USA¹⁻⁴⁾, they include ERG (End Results Group), TNCS (Third National Cancer Survey)¹⁻²⁾, SEER (Surveillance, Epidemiology, and End Results Program²⁻⁴⁾.

The Niigata Registry for Gut-Pancreatic Endocrinomas (NRGPE) started as a private collection of the literature pertaining to carcinoids around 1982, and developed into a computerized registry system based on the author's previous experience in both the experimental and clinical fields⁵⁻¹⁰⁾. This Registry has now grown into a large systemic collection of case reports and review articles dealing with endocrinocarcinomas (carcinoids and their variant endocrinomas), and endocrine tumors of the pancreas and other related organs or tissues. Articles in sixteen different languages and from sixty-five countries and regions worldwide derive from sources as diverse as the Index Medicus, Excerpta Medica, Current Contents, JCRM (Japanese Centra Revuo Medicina), as well as the numerous references cited in articles and other publications. The first largest Japanese statistical series of carcinoids was published in 1994¹¹⁾.

The purpose of the Registry is to register and put one-line all new articles published during any given year and to prepare data for researchers in the field of gut-pancreatic endocrinology that is as accurate as possible. To avoid the risk of duplication, cases where individual identification such as the age and sex of the patients were missing are excluded from the main series of the Register and every effort is made to check both the authors, the institutes where they are based and the detailed results of individual patients' laboratory examinations.

The present computerization system has been transferred from the original MS-DOS operation system¹²⁾ to the Windows version.

Financial Supports

The Registry receives financial support from a number of sources including the Japanese Ministry of Education, Science, Sports and Culture [Grant-in-Aid for Scientific Research : #01480322 & #07457268], several medical foundations as well as private donations. This support has enabled the Registry to meet its obligations, to secure the services of highly competent assistants, to gather information from worldwide sources and to get personally acquainted with many of the academics working in this field.

Contents of the Registry

As of November 2003, the Registry comprises a volume of reports amounting to 28,372 pages. The reports deal with 17,278 cases of endocrinocarcinomas (carcinoids and their variant endocrinomas), pancreatic endocrinomas as well as calcitoninomas belonging to the Carcinoid Family⁹⁾. In addition to this, the reports contain a total of 15,534 references (Table 1).

Table 1. Contents of the Niigata Registry

Endocrinomas	No. Cases	No. All Reports*	No. References**
Gut endocrinomas	14,131	21,060	10,543
1. Carcinoids & Variants ^a	5,264	9,661	6,157
2. Carcinoids & Variants ^b	7,099	9,601	4,386
3. Carcinoids/Autopsy ^a	1,768	1,798	—
Pancreatic endocrinomas***	2,730	6,524	4,558
4. Gastrinomas/ZES/Ulcerogenic syndrome ^f	381	1,268	931
5. Somatostatinomas/Inhibitory syndrome	188	352	267
6. Glucagonomas/Diabetes-Dermatitis syndrome	446	807	589
7. Insulinomas/Hypoglycemic syndrome ^g	1,141	2,888	1,791
8. Vipomas/WDHA/Diarrheogenic syndrome	247	471	379
9. PPomas	69	131	74
10. Miscellaneous endocrinomas	258	607	526
Calcitoninomas (MCT) ^a	417	788	433
Total	17,278	28,372	15,534

(Nov. 2003)

* Including duplicated case reports, cases with insufficient information and those with individual identification such as patients' age and sex.

** Including review articles.

*** Including extrapancreatic endocrinomas with corresponding hormone production.

a Japanese cases only. b Excluding Japanese cases.

MCT: medullary carcinoma of the thyroid. ZES: Zollinger-Ellison syndrome.

WDHA: watery diarrhea hypokalemia and achlorhydria.

Practical aspects of computerization : NUS analyzing system

[1] Data operation: Various keys for operating the data are available and shown in Fig. 1. It is possible to select and re-organize data, transfer the 'packaged' data to a new menu and make any further changes required.

[2] Procedures for inputting the Data :

(1) Data received is placed in one of six categories (Fig. 2). The carcinoid group including both typical and atypical varieties, and the variant group, including a range of variant endocrinocarcinomas (neuroendocrine carcinomas/NEC, endocrine small/oat cell carcinomas/SCC/OCC, endocrine cell carcinomas/ECC, etc.) are initially registered in either [1. Europe, US, and other foreign cases] or [2. Japanese cases]. The registration form is shown in Figs. 3 & 4.

(2) The primary growth sites of endocrinomas are input in an abbreviated form (Explained in the separate affiliated booklet of guidelines, e.g.: St/st for stomach, Lg/lg for lung, and Ii/ii for ileum etc.: also refer to [Addendum: Abbreviations]) The corresponding data is then input in the ways indicated.

(3) Strict data checks are conducted to avoid any possible duplication of the cases registered. If a case is registered more than once the primary registration number of this case should be the same with subnumbers ([-00], [-01], [-02], etc.) added accordingly.

Data Operation

Previous [P]
 Next [N]

Insert [I]
 Duplicate [U]
 Delete [D]

Search [S]
 Abandon [A]
 Reverse [R]

Save [S]
 Load [L]

Order change [O]
 Order release [X]

Transfer data to other menu [T]
 Change in package [P]

Fig. 1. Data Operation

NUS System: Operation

1. Europe, US, Other Foreign Cases
2. Japanese Cases
3. Combined Representative Cases
4. Autopsy Cases
5. Pancreatic endocrinomas
6. Miscellaneous Cases

1 through 4 including carcinoids
 and their variant endocrinocarcinomas.

Fig. 2. Operation for the NUS System

A	B	C	D										
Organ	No.	Au	Year										
Age	Sex	Size	mm	Multi	Site	Depth							
Weight	Mt	Gross											
Color	HxTyp	AG	AF	EM	nm/Av								
Cp/Inv	Synd	Dx											
PreDx	Mig	Op	Aut										
Post	HT	ng/ml	HI	mg/d									
ImHx:HT	Hs	Gs	NT	Sm	Vp	Mt	Gl	CT	AC	PP			
In	NSE	AFP	CEA										
											MitS0		
Lb1													
Lb2													
Tmr													
Cmp													
S/S													
Rm1													
Rm2													
Rf1													
Rf2													
Rf3													
D	E	F	G	H	I	J	K	L	M	N			

Fig. 3. Data Form Minus Data.

A	<input checked="" type="checkbox"/>	B	<input type="checkbox"/>	C	<input type="checkbox"/>	D	<input type="checkbox"/>	E	<input type="checkbox"/>														
Organ	St	No.	0038 00					Au	LattesR,US,ColumbU/S/CP,		Year	1956											
Age	28	Sex	F	Size	B	20.0mm	Multi	<input checked="" type="checkbox"/>	Site	A,	X,	Depth	sm,										
Weight	0.0g	Mt	+	Rc	Bn,Sk,Rtr,	Gross	<input checked="" type="checkbox"/>																
Color	rd-pk,	Hx	Typ	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	AG	<input checked="" type="checkbox"/>	AF	-	F,	EM	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	-	X,	nm/Av	<input checked="" type="checkbox"/>	n					
Cp	Inv	<input checked="" type="checkbox"/>	Synd	E	Dx	UG/StmU,-ChX,																	
Pre	Dx	Mlg	<input checked="" type="checkbox"/>	Op	R:Gr/Dp/RtrTmr,PtS/Pst1y,SbT/3y,PtS/DU/JU/Pst4y,							Aut	<input checked="" type="checkbox"/>										
Post	L:132m/11y/BnMt,@											HT	<input checked="" type="checkbox"/>	0.0ng/ml	HI	<input checked="" type="checkbox"/>	0.0mg/d						
Im	Hx:	HT	<input checked="" type="checkbox"/>	Hs	<input checked="" type="checkbox"/>	Gs	<input checked="" type="checkbox"/>	NT	<input checked="" type="checkbox"/>	Sm	<input checked="" type="checkbox"/>	Vp	<input checked="" type="checkbox"/>	Mt	<input checked="" type="checkbox"/>	GI	<input checked="" type="checkbox"/>	CT	<input checked="" type="checkbox"/>	AC	<input checked="" type="checkbox"/>	PP	<input checked="" type="checkbox"/>
	<input checked="" type="checkbox"/>	In	<input checked="" type="checkbox"/>	NSE	<input checked="" type="checkbox"/>	AFP	<input checked="" type="checkbox"/>	CEA	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	
	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	<input checked="" type="checkbox"/>	
Lb1	<input checked="" type="checkbox"/>																						
Lb2	<input checked="" type="checkbox"/>																						
Tmr	<input checked="" type="checkbox"/>																						
Cmp	RcDU/PyIObstr/Gr/6y,GU,RcStmU,?ZES,																						
S/S	HiAbdP,Vm,Hmt/DgBl,P:7y,																						
Rm1	Org,US,#84945,Pt:HC,BI,MltOp/5,HxG,-Mts,-MC,RcBn/11y,																						
Rm2	Typ/sm,Lst:41SCd,X,																						
Rf1	LattesR&GrossiC,Cancer 9:698-711,1956(O).																						
Rf2	Carcinoid tumors of the stomach.																						
Rf3	<input checked="" type="checkbox"/>																						
D	<input type="checkbox"/>	E	<input checked="" type="checkbox"/>	F	<input type="checkbox"/>	G	<input type="checkbox"/>	H	<input type="checkbox"/>	I	<input type="checkbox"/>	J	<input type="checkbox"/>	K	<input type="checkbox"/>	L	<input type="checkbox"/>	M	<input type="checkbox"/>	N	<input type="checkbox"/>	S:	Reported,

Fig. 4. Data Form Plus Data.

Refers to a 28-year-old female with a typical gastric carcinoid (St-0038-00), [site: antrum, tumor-size:20 mm, depth:submucosa], as reported by Lattes R & Grossi C., in Cancer 9:698-711,1956. Still alive 132 months (11 yeas) with recurrent metastases to the bone, skin and retroperitoneal region, after resection of the tumor, partial gastrectomy and subtotal gastrectomy for recurrent gastric and duodenal ulcers (Zollinger-Ellison Syndrome). Gastrin was not measured at that time.

(4) Representative cases with the subnumber [-00] are selected and transferred to [3. Combined Representative Cases] as shown in Fig. 2.

(5) The cases and items selected may be arranged in any order desired (Fig. 5) . The selected data for each case can then be displayed in a card format like the one in Fig. 4.

[3] Searching for data: Fig. 6 shows that new cases can be found by selecting any one of the featured items. Specific cases data can be either selected from or added to the series that are currently listed.

[4] Procedures for Outputting Data:

- (1) Select [Table Output] from the "Options" menu on the screen.
- (2) Select either one of the 21 representative tables in Fig. 7.
- (3) Select either one of the saved data shown in Fig. 8.

An example of an output table is shown in Fig. 9 and is obtained by selecting [1. Distribution of Gut-Pancreatic Endocrinomas.] of Fig. 7 and [1.] of the data saved in Fig. 8 with [Carcinoids: Combined cases]. Another example is shown in Fig. 10, obtained by selecting [1. Distribution of Gut-Pancreatic Endocrinomas.] from the Output Table of Fig. 7 and then selecting [1] from the data saved in the table for pancreatic endocrinomas with the key [Pancreatic endocrinomas]. Some aspects of metastases are shown as being limited to the rectal (Rc) endocrinocarcinomas in

Figs. 11 and 12. The cases in Fig. 11 are those in which the tumor-sizes are judged to belong to the ABC size-categories, and the cases in Fig. 12 are those in which the tumor-sizes are recorded on a millimeter scale. As an example, the syndromes in pancreatic endocrinomas are shown together with either the corresponding organs or growth sites of origin and the incidence rates of the syndromes (Fig. 13).

Year	Organ	Number	Abn?	Age	Sex	Author	Size	Syn	Mar/O
2002	Ap	0834	00	19	F	KocaAM,Blg EtterbeekIxellesHISCntH/GES,	13	-	T
2002	Ap	0838	00	58	F	GarinL,Fr,CentreEMarquis/OncM/Gy/AP.,	25	-	A
2003	Ch	0003	00	19	M	VolpeCM,US,NYStateU/S/BioMSci,	10	-	T
2000	Co	0232	00	30	F	JainS,UK,NottinghamCityH/S/P,	10	-	T
1997	Dd	0337	00	65	M	FresnoMF,Sp,OviedoU/CovadongaU/AP,	30	-	A
2002	Es	0063	00	48	M	HoangMP,Albores-SaavedraJ,US,TxsU/SouthWestMC/P,AFIP/Hep&GIPathol,	35	-	T
2001	Gb	0128	00	38	M	SinkraPA,US,UTSouthWestMC/P,APIP/H&GIP,	14	-	T
2001	Hp	0119	00	66	F	FurrerJ, Sws,ZurichU/MedCl/M/R,	70	+	A
2001	Ic	0060	00	47	F	SimanekV,Cze,PlzenLochotin/S/P/M,	23	-	T
2002	Ii	0989	00	59	F	ShehataB,US,EmoryU/EglestonChildren'sH/OhioMedColl/P,	23	+	T
2001	Ii	0947	00	45	F	PaIT,FrCan,RsC/WomensHealthSunnyBrook&MomensCol/EdwardHerriotH/GenetU,	30	+	T
2002	Kd	0503	00	68	M	GunesA,Trk,NonuU/Uro/P,	110	-	T
2001	Lg	1381	00	74	F	MinKW,US,DeaconessH/P,	32	-	A
2003	Lg	1438	00	27	F	LoliP,It,MilanNiguardaH/End/NucM/ThS/R/P/S,BustoArsizioH/S/NucM,	20	=	T
2001	Lx	0524	00	51	F	KeberleM,Ger,Wurzburg,?Inst,	15	-	A
2002	Ov	0264	00	54	F	TargaL,It,EsteCivileH/M/CardVsc,	20	+	T
1992	Rc	0525	00	50	M	McNeelyB,Can,BritishColumbiaU/P/S,VancouverGnH/,	2	-	T
2002	St	0522	00	52	M	AdhikariD,US,NorthShoreU/NYU/P/S/M,	25	-	A
2003	Th	0237	00	42	M	GibriF,US,NIH/DgDis/DM/Kd/ThOnc/MolPathg/SNeuro/NucM/DxR/,	80	-	T
2001	Ts	0521	00	68	M	StingerAJ,US,SouthernCalPermanenteMedGr/U/P,CalifU/P/LbM,	14.5	-	T
2002	Ub	0008	00	75	F	AkimovOV,Rs,SmolenskRegionalPatholInst,	15	-	T

Fig. 5. A Series of Cases Selected and Arranged in a Preferred Order.

It is possible to make a further selection of any case for evaluation purposes or for printing out in card form.

Searching Cases

New Cases

Select Cases from the present series available

Additional Cases into the present series available

Release Searching

Reverse Data against the present series available

Fig. 6. Searching Procedures for Specific Cases. In: [OPTION (O)]

Table Output	
1.	Distribution of Gut-Pancreatic Endocrinomas
2.	M/F ratio
3.	Age: Average, Distribution & Range
4.	Signs & Symptoms
5.	Metastases (1)
6.	Metastases (2)
7.	Silver Impregnation
8.	6-HT Activity
9.	The Syndromes (1)
10.	The Syndromes
11.	Size distribution
12.	Size & Metastases (1)
13.	Size & Metastases (2)
14.	Size & Metastases (3)
15.	Average Tumor-Size (mm)
16.	Histologic Types
17.	Histologic Types & Metastases
18.	Depth of Invasion
19.	Depth & Invasion & Metastases
20.	Immunohistochemistry
21.	Chronological Analysis of Reported Cases

Fig. 7. Basic Table Forms for Printing Data Out.

1	12152CdAll/[T+A+V]/031030,
2	274CCd/[T+A]/031010,
3	24[+Mt]/DgsmCd/Ex:sm/Sz>0/[T+A]/0.1-5mm/Frm#14/031011,
4	Rc1018Cd/In:R/Ex:NB/X/ND/000/[T+A+V]/031020,
5	6516Cd/In:R/Ex:NB/X/ND/000/[T+A+V]/031020,
6	1730RCd/[T+A+V]/031007,
7	1651RCd/[T+A]/031007,
8	1611RCd/[T]/031007,
9	7056AllDgCdG/[T+A]/031014,
10	7563AllDgCd/[T+A+V]/031014,
11	5YS/823RCd[T+A]/In:R/Ex[NB,Lst,X,ND,000/O/?m/Dbl/?D?L?m/In[Dbl
12	773smRCd/Ex:sm/Sz>0/[759T+9A+5V]/031009,
13	1679smCd/Ex:sm/Sz>0/[1576T+46A+57V]/031011,
14	1599DgsmCd/Ex:sm/Sz>0/[1565T+34A]/031011,
15	1078AllVariatG/[V]/031014,
16	5YS/5889AllCd[T+A+V]/In:R/Ex[NB,Lst,X,ND,000/O/?m/Dbl/?D?L/I
17	5YS/5886AllCd[T+A]/In:R/Ex[NB,Lst,X,ND,000/O/?m/Dbl/?D?L/In[I
18	5YS/4611AllCd[T]/In:R/Ex[NB,Lst,X,ND,000/O/?m/Dbl/?D?L/In[Dbl
19	5YS/775AllCd[A]/In:R/Ex[NB,Lst,X,ND,000/O/?m/Dbl/?D?L/In[Dbl.C
20	11838Cd/[T+A+V]/030714,[Ex:hp-0106→0118-03/st-0846→ 0849-01][dd-0456-00→dd-0248-00/rc-0312→0080-01]030811],

[Select the number / Cancel]

Fig. 8. Data Saved

1 . Distribution of Carcinoids

03/11/05

Organ	Number	Dig.only(%)	All(%)
Es	126	1.7	1.0
St	1382	18.3	11.4
Dd	948	12.5	7.8
Jj	124	1.6	1.0
li	1081	14.3	8.9
lj	119	1.6	1.0
lc	94	1.2	0.8
Ap	1047	13.8	8.6
Co	345	4.6	2.8
Rc	1730	22.9	14.2
Gb	151	2.0	1.2
Hp	155	2.0	1.3
Ch	84	1.1	0.7
Pn	130	1.7	1.1
Dg	47	0.6	0.4
SubT	7563	100.0	62.2
Lg	2367		19.5
Th	540		4.4
Br	333		2.7
Ov	368		3.0
Lx	285		2.3
Cx	222		1.8
Kd	77		0.6
Ts	72		0.6
Ea	80		0.7
Ub	83		0.7
M	162		1.3
SubT	4589		37.8
Total	12152		100.0
Jj+li+lj	1324	17.5	10.9

1 - 12152CdAll/[T+A+V]//031030,

Fig. 9.

Distribution of Primary Growth Sites
for Endocrinocarcinomas.

1 . Distribution of Endocrinomas

03/12/03

Organ	Number	(%)	Panc.only	(%)
ZE	381	12.1	235 (61.7%)	
Sm	188	6.0	88 (46.8%)	
Gl	446	14.2	434 (97.3%)	
In	1141	36.3	1114 (97.6%)	
Vp	247	7.8	184 (74.5%)	
PP	69	2.2	64 (92.8%)	
Cd	115	3.7	109 (94.8%)	
Ms	143	4.5	133 (93.0%)	
Md	417	13.3	0 (0.0%)	
Total	3147	100.0	2361 (75.0%)	

Fig. 10. Distribution of Primary Growth Sites
for Pancreatic Endocrinomas.

13 . Size & Metastases (2)

Organ : Rc

03/12/02

	A	B	C	D	E	F	G	H	I	SubT	X	Total
Number	930	297	87	38	40	39	22	6	4	1463	267	1730
N	53	83	34	27	20	23	15	3	4	262	52	314
	5.7	27.9	39.1	71.1	50.0	59.0	68.2	50.0	100.0	17.9	19.5	18.2
H	34	50	44	28	20	20	11	2	1	210	73	283
	3.7	16.8	50.6	73.7	50.0	51.3	50.0	33.3	25.0	14.4	27.3	16.4
L		6	2	3	5	4	3	1	1	25	8	33
		2.0	2.3	7.9	12.5	10.3	13.6	16.7	25.0	1.7	3.0	1.9
Bn	5	4	5	4	4	1	3	1		27	18	45
	0.5	1.3	5.7	10.5	10.0	2.6	13.6	16.7		1.8	6.7	2.6
Prt		7	8	1	4	4	2			26	12	38
		2.4	9.2	2.6	10.0	10.3	9.1			1.8	4.5	2.2
Adr	1		3	1	3		1	1		10	3	13
	0.1		3.4	2.6	7.5		4.5	16.7		0.7	1.1	0.8
P		3	5	1	2		1	1		13	2	15
		1.0	5.7	2.6	5.0		4.5	16.7		0.9	0.7	0.9
-	279	66	15	2	4	4	2	1		373	29	402
	30.0	22.2	17.2	5.3	10.0	10.3	9.1	16.7		25.5	10.9	23.2
X	569	114	9	1	7	2	1	2		705	130	835
	61.2	38.4	10.3	2.6	17.5	5.1	4.5	33.3		48.2	48.7	48.3
+	81	116	63	35	29	33	19	3	4	383	107	490
	8.7	39.1	72.4	92.1	72.5	84.6	86.4	50.0	100.0	26.2	40.1	28.3
?	1	1								2	1	3
	0.1	0.3								0.1	0.4	0.2

1 - 12152CdAll/[T+A+V]//031030,

Fig. 11. Rectal Endocrinocarcinomas: Relationship between Tumor-Size and Sites of Metastasis.

T: Typical carcinoid. A: Atypical carcinoid. V: Variant endocrinoma.

14 . Size & Metastases (3)

Organ : Rc

	A		B		C	D	E	F	G	H	I	SubT	Avr. Size	X	Total
	~5	~10	~15	~20											
Total	864		285												
Mt(+)	77		114												
(%)	8.9		40.0												
Total	338	526	196	89	85	37	39	25	19	5	2	1361	14.7	369	1730
Mt(+)	11	66	69	45	62	34	29	21	16	2	2	357	28.6	133	490
(%)	3.3	12.5	35.2	50.6	72.9	91.9	74.4	84.0	84.2	40.0	100.0	26.2		36.0	28.3

1 - 12152CdAll/[T+A+V]//031030,

Fig. 12. Rectal Endocrinocarcinomas: Relationship between Tumor-Size on a Millimeter Scale and Rates of Metastases.

9. The Syndromes (1)

03/12/03

Organ	Number	(+)	%	(X)	(?)	(-)	(=)
ZE	381	319	83.7	1	32	24	5
Sm	188	21	11.2	0	10	126	31
GI	446	237	53.1	20	0	122	65
In	1141	997	87.4	42	43	28	31
Vp	247	214	86.6	0	20	7	6
PP	69	0	0.0	8	10	42	9
Cd	115	21	18.3	58	3	30	3
Ms	143	16	11.2	10	6	99	10
Md	417	1	0.2	380	0	29	7
Total	3147	1826	58.0	519	124	507	167

4 - 3147Cases/00/031203,

Fig. 13. The Incidence and Rate of Syndromes for Pancreatic Endocrinomas

Characteristic features of the Registry

1) The Registry retains only those cases that have been published in authorized journals or reported at official congresses. Some of the cases are supplemented by correspondence between the present writer and the authors of the corresponding articles.

2) Each of the approximately 17,000 cases and 28,000 references has been individually evaluated and systematically categorized.

3) The data that has been collected worldwide (from the aforementioned sixty-five countries and in sixteen languages) can also be supplied worldwide. Every effort is made to ensure that researchers are supplied with all available information stored in the Registry database.

4) More than 42% of the data stored in the 'Gut-Endocrinocarcinoma Series' is taken from Japanese publications

5) One can carry out a comparative evaluation of gut-pancreatic endocrinomas in Japan and other countries or regions by analyzing various factors such as genetic, socioeconomic or geographical statistics. The evaluation reveals that between the Japanese and non-Japanese, there is a statistically significant difference in the primary growth sites of carcinoids in the appendix, small intestine, rectum and stomach¹¹⁾.

Publications based on NUS analyzing system

Table 1 shows that during the past 15 years, from 1989 to 2003, approximately 17,300 cases of endocrinocarcinomas and pancreatic endocrinomas along with calcitoninomas have been registered. Following the occasional re-evaluation of individual cases and their corresponding data in the category of 'primary growth sites of origin', they have been frequently published (Table 2).

Table 2. List of publications based on the data from the Niigata Registry

Subjects	Remarks
1889-1993	
Malignancy of carcinoids.	A comparison of submucosal lesions of an identical size between GI carcinoids & gastric carcinomas ¹³⁾
Carcinoid somatostatinoma / duodenum.	Concept of carcinoids & pancreatic endocrinomas ¹⁴⁾
Gut-pancreatic endocrinomas.	Statistics of 4625 cases ¹⁵⁾
1994-1997	
Pancreatic endocrinomas.	Statistics of 1857 cases ¹⁶⁾
PPomas.	Analysis of 58 cases & their characteristics ¹⁷⁾
Carcinoids: Japanese series.	Analysis of 3126 reported & 11810 autopsy cases ¹¹⁾
Carcinoids of the appendix.	A case report & statistics of 142 Japanese cases ¹⁸⁾
Goblet cell carcinoids of the appendix.	Statistics of 170 cases compared to 456 ordinary type appendiceal carcinoids ¹⁹⁾
Composite patterns of gastric carcinoids.	Two gastric carcinoids & classification of composite patterns ²⁰⁾
Duodenal carcinoids.	Statistics of 635 cases ²¹⁾
Hepatobiliary pancreatic carcinoids.	Statistics of 266 cases ²²⁾
Rectal carcinoids.	Statistics of 1271 cases ²³⁾
History of carcinoid research.	Including the period since 1837 before proposal of the concept of "Carcinoid" in 1907 ²⁴⁾
Gastric carcinoids.	Statistics of 1094 cases ²⁵⁾
Carcinoids of the small intestine.	Statistics of 1102 cases ²⁶⁾
1998-1999	
Carcinoids with metastases.	Statistics of 2001 cases ²⁷⁾
Esophageal endocrinomas.	Statistics of 100 cases: 28 typical & 78 atypical varieties ²⁸⁾
Gastrinomas/Zollinger-Ellison syndrome.	Statistics of 359 Japanese cases ²⁹⁾
Colonic & ileocecal carcinoids.	Statistics of 363 cases ³⁰⁾
Glucagonomas/diabetico-dermatogenic syndrome/DDS.	Statistics of 407 cases ³¹⁾
Insulinomas/hypoglycemic syndrome.	Statistics of 1085 Japanese cases ³²⁾
Vipomas/diarrheogenic syndrome.	Statistics of 241 cases ³³⁾
Classification of gut endocrinomas.	Classification problems of carcinoids & their variant endocrinomas ³⁴⁾
Somatostatinomas/ inhibitory syndrome.	Statistics of 173 cases ³⁵⁾
Carcinoid syndrome.	Statistics of 748 cases ³⁶⁾
Lung/ bronchial carcinoids.	Statistics of 1875 cases/ typical & atypical varieties ³⁷⁾
Mediastinal/ thymic carcinoids.	Statistics of 432 cases ³⁸⁾
2000-2003	
Ovarian carcinoids.	Statistics of 329 cases ³⁹⁾
Carcinoids of the uterine cervix.	Statistics of 205 cases ⁴⁰⁾
Mammary carcinoids.	Statistics of 310 cases/ typical & variant ⁴¹⁾
Laryngeal endocrinomas.	Statistics of 278 cases/ carcinoid & variant groups ⁴²⁾
Hepatic carcinoids.	Statistics of 126 cases/ carcinoid 6 variant groups ⁴³⁾
Endocrinomas/ gallbladder.	Statistics of 138 cases/ carcinoid & variant groups ⁴⁴⁾
Duodenal endocrinomas.	Statistics of 927 cases/ carcinoid & variant groups ⁴⁵⁾
Endocrinocarcinomas.	Statistics of 11842 cases/ Carcinoid & variant group ⁴⁶⁾

The current research activities in the field of gut-pancreatic endocrinology

The trend of international research in the field of gut-pancreatic endocrinology since 1970, is shown chronologically in the graph (Fig. 14). This graph illustrates the increasing volume of the recent research in three areas:

- A. In the total number of reports including review articles and case reports that are either duplicated or insufficiently well-documented.
- B. In the total number of references.
- C. In the total number of effective case study reports.

Occasional publications include statistical analyses of national trends, based on a considerable number of cases of gut-pancreatic endocrinomas. In addition to the Japanese series stored in the Niigata Registry (carcinoids and their variant endocrinomas and pancreatic endocrinomas as shown in Table 2^{11,29,32}), representative works were produced by the following: (1) the End Cooperative Oncology Group (ECOG: carcinoids of all sites, USA)⁴⁷, (3) the Z-E Tumor Registry (Zollinger-Ellison syndrome/gastrinomas, USA.)⁴⁸, (4) the Connecticut Tumor Registry (carcinoids of the colon, USA)⁴⁹, (5) the Utah Cancer Registry (carcinoids of all sites, USA)⁵⁰, (6) the Alberta Cancer Registry (carcinoids of the colon, Canada)⁶¹, (7) the Danish Cancer Registry (carcinoids of all sites, Denmark)^{52,53}, and (8) SEER (Surveillance, Epidemiology, and End Results Program, carcinoids of all sites, USA)².

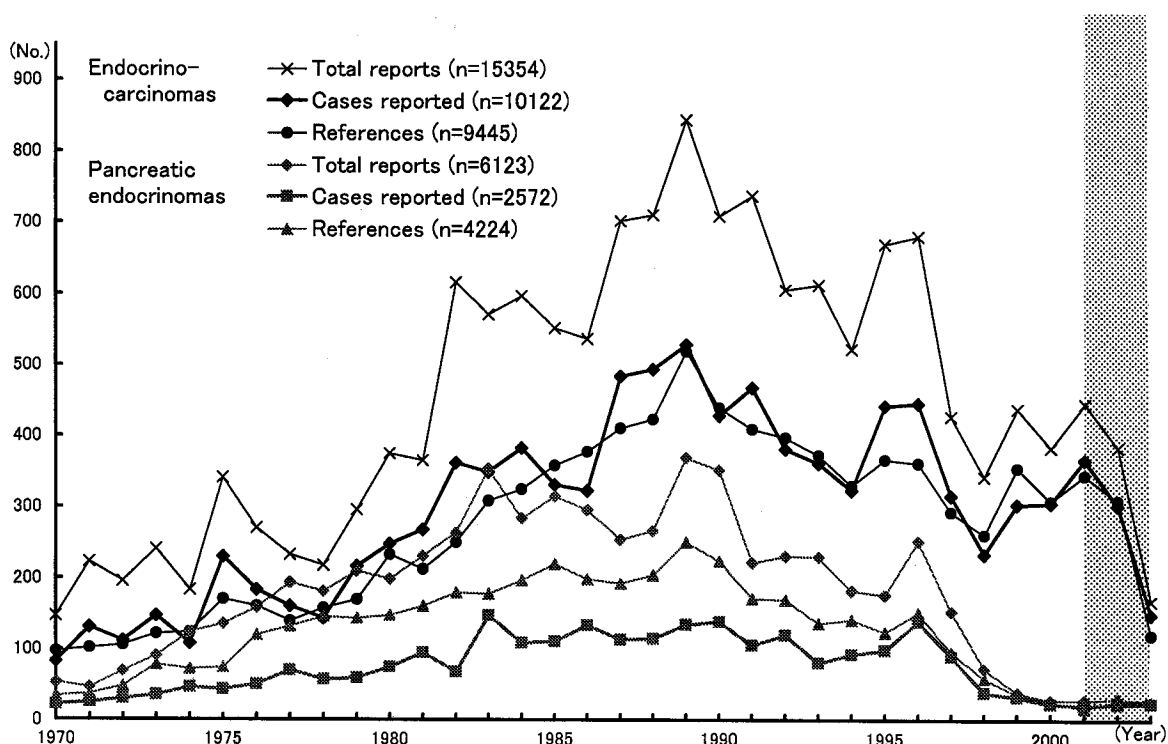


Fig. 14. Chronological Variations of the Number of Reports, Cases and References Published for Both Series of Endocrino-carcinomas and Pancreatic Endocrinomas.

Comments

The Niigata Registry, which had initially been under the sole supervision of the present writer, was donated to Niigata University School of Medicine, with its accompanying NUS System and a large body of literature. It is now housed in a special section of the Asahimachi Branch of the Niigata University Library. It is hoped that this guide to both the registry and the NUS operating system, will be of assistance to those working in the field. It is important that the Registry continues its work of accumulating annually new data from around the world.

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[ADDENDUM: Abbreviations] :

[A] Sites/ Organs: Syndromes

1) Carcinoid Series: Foreign/ Japanese

Es/es = Esophagus	Gb/gb Gallbladder	Lx/lx = Larynx
St/st = Stomach	Hp/hp= Liver/Heptic	Kd/kd= Kidney
Dd/dd = Duodenum	Ch/ch= Cheledochus	Ts/ts= Testicle
Ij/ij = Jejunum	Pn/pn= Pancreas	Ea/ea= Middle ear
Ii/ii = Ileum	Dg/dg= Digestiveorgan	Ub/ub= Urinary bladder
Ij/ij= Jejunoileum	Lg/lg = Lung	M/m = Micellaneous sites
Ic/ic= Ileoecum	Br/br = Breast	Th/th = Thymus/ Mediastinum
Co/co = Colon	Ov/ov= Ovary	
Rc/rc = Rectum	Cx/cx= Cervix (uterine)	

2) Pancreatic Endocrinomas

ZE : Zollinger-Ellison Syndrome/ Gastrinomas (Japanese only)
In : Insulinomas (Japanese only).
Sm : Somatostatinomas
Vp : Vipomas
Gl : Glucagonomas.
PP : PPomas
Cd : Pancreatic carcinoids.
Md : Medullary carcinomas of the thyroid (MCT:Japanese only)
Ms : Miscellaneous tumors.

[B] Countries/ Regions

Arg,	Argentina:	Ind,	India	Rs,	Russia
AUS,	Australia:	Ir,	Ireland	Rum,	Rumania
Aus,	Austria	Isr,	Israel	SAr,	Saudi Arabia
Blg,	Belgium	It,	Italy	SAr,	South Africa
Blv	Bolivia	Jam,	Jamaica	Slv,	Slovenia
Brz,	Brazil	Jp,	Japan	Sneg,	Senegal
Bul,	Bulgaria	Kor,	Korea	Sng,	Singapol
Can,	Canada	Kwt,	Kuwait	Sp,	Spain
Chl,	Chile	Leb,	Lebanon	Swe,	Sweden
Chn,	China	Mal,	Malacia	Sws,	Swis
CR,	Costa Rica	Moro,	Morocco,	Tha,	Thailand
Cro	Croatia	MX/Mx,	Mexico	Trk,	Turky
Cze,	Czechoslovakia	Ngr,	Naigeria	Tun,	Tunisia
Dnm,	Denmark	NLD,	Netherland	Twn,	Taiwan
Fr,	France	Nrw,	Norway	UK,	England
FNL,	Finland	NZ,	New Zealand	Uk,	Ukraine
Egy,	Egypt	Oma,	Oman	Urg,	Urguay
Ger,	Germany	Pak,	Pakistan	US,	United States
Gr,	Greek	Pol,	Poland	Yug,	Yugoslavia
Hng,	Hong Kong	Prt,	Portugal	Zim,	Zimbabwe
Hun,	Hungary	PRco,	Puerto Rico		
ICL,	Iceland	Vnz,	Venezuera		

[C] Japan : Prefecture / Cities / Districts

Ach	Aichi	Mzk	Miyazaki	Shg	Shiga
Akt	Akita	Ngs	Nagasaki	Shk	Shikoku
Aom	Aomori	Ngt	Niigata	Shn	Shinshu
Asa	Asahigawa.	Ngy	Nagoya	Shz	Shizuoka
Chb	Chiba	Nra	Nara	Spp	Sapporo
Ehm	Ehime	Oht	Ohita	Stm	Saitama
Fki	Fukui	Oky	Okayama	Tch	Tochigi
Fks	Fks	Kch	Kouchi	Ttr	Tottori
Fuk	Fukuoka	Kgs	Kagoshima	Tks	Tokushima
Gif	Gifu	Kgw	Kagawa	Tsk	Tsukuba
Gnm	Gunma	Kmt	Kumamoto	Tym	Toyama
Hsk	Hirosaki	Kng	Kanagawa	Wky	Wakayama
Hkk	Hokkaido	Knz	Kanazawa	Ych	Yamaguchi
Hmt	Hamamatsu	Krm	Kurume	Ygt	Yamagata
Hrs	Hiroshima	Kws	Kawasaki	Ykh	Yokohama
Hyg	Hyogo	Kys	Kyushu	Yks	Yokosuga
Iwt	Iwate	Kyt	Kyoto	Ymn	Yamanashi
Kbe	Koube	Osk	Osaka		
Mie	Mie	Ryk	Ryukyuu		

腸膵内分泌腫瘍の登録・保存・分析・供給のための新潟システム：Window型変換

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要 旨

Niigata Registry for Gut-Pancreatic Endocrinomas (NRGPE) はいわゆる消化管ホルモンを産生・分泌する消化器内分泌癌 (Endocrinocarcinomas: カルチノイド群・類縁腫瘍群) および膵内分泌腫瘍の登録・保存・分析・供給を目的とするシステムである。その内容は内外において報告された個人識別の可能な症例のほか、本邦剖検症例、統計分析の対象外ではあるが内外の個人識別不可能な群別報告症例、綜説などが収録・保管されている。本邦を含む65カ国より渉猟された16言語による文献約15,500部、症例17,000例、総件数28,000件が系統的にデータベース化されている。基本的分析表は21種類あり、それらの組み合わせにより、内外の研究者の要望に応じて必要な資料を供給することが出来る。消化器内分泌癌では本邦症例が約42%を占め、外国症例との統計的有意差検定も可能である。本来は個人の収集資料であったが、膨大な資料と独特なデータベースであるため、一般公開を建前として、1999年3月に新潟大学に寄贈し、現在、大学図書館医学部旭町分館特別室に保管されている。

本著では、このRegistryの機能の概略、内容、公表された分析結果、資料抽出・解析の実際等を解説し、今後この専門領域に従事する内外の研究者に資料を提供する方策を示した。

キーワード

腸膵内分泌腫瘍新潟登録、NUSシステム、カルチノイド、内分泌癌、神経内分泌癌、内分泌細胞癌