Breast cancer and screening in persons with an intellectual disability living in institutions in France

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Abstract

Background   We aimed to describe, among a population of women with intellectual disabilities (ID) living in institutions in France, the characteristics in whom breast cancer (BC) was diagnosed and of those who participated in BC screening.

Methods   Study was performed in 2009 among a random, representative sample of women with ID living in institutions in France. Participants answered a questionnaire either directly by themselves, or with the help of an intermediary.

Results   In total, 978 women with ID aged over 18 years were included, and 14 were diagnosed with BC. The incidence observed in this sample of women with ID is similar to that of the general population (standardised incidence ratio, SIR 0.857, 95% confidence interval (CI) 0.42–1.53). Average age at diagnosis was 47.8 years, and the risk of developing BC before the age of 50 was 2.03% (0.4–3.66). This risk was not significantly different from that of the general population (2.4%, 1.0–3.78). Obesity was almost twice as frequent in women who had BC as compared to those without BC (43% vs. 22.5%, $P = 0.0196$). Among the 310 women aged >50 years

and eligible for the national BC screening programme, 238 (77%) had already had at least one mammogram, and 199 had had it within the previous 2 years. Adherence to the screening programme was 64.2% (199/310) in the participating institutions. This rate was slightly higher than the national average of 62% for the same period.

Conclusions   The results of this study show that BC is equally as frequent among women with ID living in institutions as in the general population, and occurs at around the same age. Obesity was significantly more frequent among women in whom BC was diagnosed in our study. Participation in BC screening is slightly higher among women with ID living in institutions than among the general population.

Keywords   breast cancer, France, institution, intellectual disability, obesity, screening

Introduction

The incidence and mortality of cancer in persons with intellectual disabilities (ID) are estimated to be comparable to those of the general population (Patja *et al.* 2001; Sullivan *et al.* 2004; Kiani *et al.* 2010). However, the distribution of cancers by affected organ is somewhat different than in the general population and can vary according to the type and

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origin of the ID (Satgé *et al.* 2007). Epidemiological studies of incidence (Patja *et al.* 2001; Sullivan *et al.* 2004) and mortality (Kiani *et al.* 2010), as well as reported experiences from institutions (Jancar & Jancar 1977; Evenhuis *et al.* 1996), suggest that persons with ID develop cancer at approximately the same rate as persons without ID. This is the case for breast cancer (BC). However, the frequency of BC is not homogeneous, since certain genetic conditions associated with ID are associated with an increased risk of BC, such as Cowden syndrome (Brownstein *et al.* 1978), neurofibromatosis 1 (Sharif *et al.* 2007) or Saethre–Chotzen syndrome (Sahlin *et al.* 2007), whereas other conditions are associated with a decreased risk of BC, e.g. Down syndrome (Satgé *et al.* 2001) or Fragile X syndrome (Farach *et al.* 2013).

In France, with an estimated 48 763 new cases in 2012, BC remains the most common cancer in women and is also the primary cause of cancer-related death in women, with an estimated 11 900 deaths in 2012 (Binder-Foucard *et al.* 2014). The prognosis of BC has improved strikingly over the past 20 years, and standardised net survival is among the highest of all cancer types, at 85% at 5 years, and 76% at 10 years (Cowppli-Bony *et al.* 2016). Breast tumours are being discovered at ever earlier stages thanks to screening programmes (Foca *et al.* 2013), and considerable progress has been made in recent years in terms of therapeutic options (Anampa *et al.* 2015).

Organised BC screening is implemented throughout the whole of France since 2004 for women aged 50 to 74 years. Eligible women are invited to attend a screening mammogram every 2 years. The screening programme aims to provide equal access to screening for women everywhere in France, and each participant is guaranteed to benefit from the same quality of care. The rate of participation in national screening for the period 2009–2010 was 52% (Rogel *et al.* 2012). However, women also have the possibility to undergo screening mammograms on prescription from a health professional (individual screening). The contribution of individual screening among the target population was estimated to be 10% in 2009 (Hirtzlin *et al.* 2012), thus yielding an overall rate of screening participation of 62% (Haute Autorité de Santé 2011).

Previous studies of BC in women with ID mainly addressed the question of screening (e.g. the rate of participation, access to screening, barriers to participation, awareness of screening among women with ID). Data are sparse concerning the frequency and characteristics of BC in this population. Furthermore, to the best of our knowledge, there are no current data available regarding the frequency of BC at a national level among women with ID living in institutions.

Therefore, the aim of this study was to describe the characteristics of women with ID living in institutions, those diagnosed with BC and those who participated in BC screening, based on the national 'disabled persons healthcare survey' (Handicap Santé en Institution), performed by the French National Institute of Statistics and Economic Studies (Institut National de la Statistique et des Etudes Economiques, INSEE) in 2009.

Methods

Disabled persons healthcare survey

First performed in 2009, the disabled persons healthcare survey – institutional section was jointly organised by the INSEE and the research, studies and statistical analysis directorate (Direction de la recherche, des études, de l'évaluation et des statistiques, Drees) of the National Ministry for Health.

The disabled persons healthcare survey was performed among a random, representative sample of institutions and residents in various establishment in metropolitan France and French overseas territories. The participating institutions included institutions for handicapped adults, institutions for persons with psychiatric disorders, institutions for elderly subjects, as well as residential centres for social reinsertion. From among the database of health establishments accredited by the National Ministry for Health, which includes approximately 12 000 institutions, 1519 institutions were randomly selected. A second random selection was performed on site in each institution by the survey agent to randomly identify six to eight residents per institution to participate in the survey. No resident present in any institution was excluded on the basis that they were unable to participate in the interview.

In total, 9104 persons were interviewed in 1519 randomly selected institutions within the disabled

persons healthcare survey – institutional section. The full methodology used for this survey has previously been described elsewhere (Bourgarel *et al.* 2015).

Inclusion criteria

Our study concerned adult women (aged >18 years) with an ID and living in an institution, who participated in the disabled persons healthcare survey. Subjects were considered to have ID if they selected 'mental retardation' among a list of 12 psychological disorders proposed in the questionnaire.

We selected three types of residential institutions that specialise in the management of persons with ID, namely specialised residential institutions, medicalised residential institutions and community-style residential accommodation. Specialised residential institutions are medico-social establishments in which adults who live there require full assistance with daily living and whose medical status requires medical surveillance and regular care. Medicalised residential institutions cater for persons with disabilities who are unable to engage in professional activity and who require assistance for the majority of essential tasks of daily living, as well as medical surveillance and regular care. Community-style residential facilities are designed for persons with disabilities who cannot (or can no longer) work, but who nonetheless possess a certain degree of physical and/or intellectual autonomy. This type of residential setting aims to provide accommodation at night and at the weekend for persons with ID who are working in specialised structures, in special-needs-adapted businesses or in an ordinary professional setting. Nursing homes for elderly subjects were excluded in order to exclude patients with ageing-related cognitive disorders. For the purposes of clarity, in the presentation of our results, we grouped the participating institutions into two categories, namely specialised residential institutions (the first two types) and community-style residential facilities (the third type).

In total, 978 women were included in this analysis, constituting a representative sample of women with ID living in institutions in France.

Data collection – questionnaire

Data collection was performed on site in each institution in person-to-person interviews, using a dedicated questionnaire (Bouvier 2011). For participants who were randomly selected to participate, but unable to be directly interviewed, data collection was performed with the help of a caregiver or another healthcare professional from the institution who knew the participants, or as a last resort, with the help of one of the participant's close relatives designated by the institution.

The data analysed here are taken from the disabled persons healthcare survey individual questionnaire, which comprised over 100 pages dealing with health, functional status (disease, incapacities, limitations), the different types of assistance received (technical, personal and financial), diet, the person's environment, level of education, employment (if any), resources, as well as certain factors of social participation and environmental factors.

The survey, which was conducted at the national level by the National Institute of Statistics and Economic Studies (Institut National de la Statistique et des Etudes Economiques, INSEE), received ethics committee approval and was also approved by the national authority for the protection of privacy and personal data (Commission Nationale de l'Informatique et des Libertés, CNIL) under the number 1259545-July 2008. The consent of each participant was obtained either from the person him-/herself where possible, or from their next-of-kin. The manager determined whether the adults with ID had the capacity to provide consent. The extraction of anonymised data was approved by the Centre Maurice Halwachs (N° 7331-August 20103).

Statistical methods

Quantitative data are presented as mean ± standard deviation and qualitative data as number (percentage). Characteristics were compared using the Chi square or Student *t* test as appropriate. We calculated the expected number of incident BC cancers between 1999 and 2009 in our study cohort using indirect standardisation, based on the assumption that crude rates per age category and per year for BC in this population would be similar to those of the general French population, taken from the FRANCIM database (Binder-Foucard *et al.* 2014).

The number of persons at risk in each age category was estimated for each year based on the cohort, for whom the age was known in 2009 and thus back-calculated for the previous years. The standardised incidence ratio (SIR) was calculated by dividing the number of BC cases observed in the period 1999–2009 by the number of expected cases over the same period. The 95% confidence interval (CI) for the SIR was also calculated. The cumulative risk of having BC before the age of 50 was calculated for the French general population, and for the population of women with ID. This risk corresponds to an individual's risk of developing cancer up to a certain age, if there were no other causes of death. It is estimated as the cumulative incidence rate (the sum of specific incidence rates from birth to age 50). It is expressed as a percentage with 95%CI. All analyses were performed using SAS version 9.3 (SAS Institute Inc., Cary, NC, USA).

Results

In total, 99.5% of specialised residential facilities in which adults with ID live participated in the disabled persons healthcare survey. Among these, 93.8% of persons randomly selected within the institutions to participate actually completed the questionnaire.

Study population

Among the 978 women aged >18 years with ID and living in residential facilities for adults with disabilities, half (49.5%) were living in specialised institutions, and the other half (50.5%) in community-style residential facilities. Average age was 42.8 years (±11.9), and almost a third (31.7%) were aged over 50 years. The distribution of the number of person-years per age category in the cohort of women with ID for the period 1999 and 2009 showed an over-representation of the younger age groups and an under-representation of the most advanced age groups as compared to the general female population (Table 1). The majority of women with ID had a legal guardian (89.2%). There were 209 women (21.4%) employed at the time of the study, and 18 (1.8%) were in a relationship. Over half (52.8%) had normal morphological characteristics, with a body mass index (BMI) <25, while 28.4% were considered overweight (25 ≤ BMI < 30), 17.3% were considered obese (30 ≤ BMI < 40) and 1.5% were

Table 1 Distribution of person-years per age category in the general French population and in the study population of women with intellectual disabilities living in institutions, 1999 to 2009

Age category	General population		Women with ID	
	Person-years	Proportion	Person-years	Proportion
20_24	21 256 233	8%	984	10%
25_29	21 516 294	8%	1110	11%
30_34	22 808 145	9%	1308	13%
35_39	24 124 882	9%	1495	15%
40_44	24 192 322	9%	1556	16%
45_49	23 776 448	9%	1464	15%
50_54	23 307 777	9%	1094	11%
55_59	20 591 684	8%	571	6%
60_64	16 757 617	6%	228	2%
65_69	15 309 990	6%	67	1%
70_74	15 413 132	6%	15	0%
75_79	14 370 945	5%	2	0%
80_84	10 667 747	4%	5	0%
85sup	10 666 673	4%	4	0%
Total	264 759 889	100%	9903	100%

morbidly obese (BMI ≥ 40) (Table 2). In all, 20 cancers were diagnosed in the study population, of which 14 were BC, one colorectal cancer, one uterine cancer and four with unspecified localisation.

Breast cancer

Among those with BC, eight (57.2%) were aged under 50 years of age at diagnosis. Average age at diagnosis was 47.8 years (±9.6) (Table 3).

• *Comparison between ID and non ID population*

The cumulative risk of developing BC before the age of 50 in the ID population was 2.03% (95%CI 0.4%, 3.66%). This risk was not significantly different from that of the general population (2.4%; 95%CI 1.0%–3.78%). Among the 14 cases of BC, 11 were incident cases declared during the period 1999–2009, with possibly one additional case in a woman of 62 years of age who did not know the date of her diagnosis (Table 3). The expected number of cases for the same period was 12.8, based on the national incidence rate per age category and per year (SIR 0.857, 95%CI 0.42–1.53, *P* = 0.53).

Table 2 Distribution of women with intellectual disabilities aged <50 years vs. ≥ 50 years according to body mass index

Age	BMI < 25	25 ≤ BMI < 30	30 ≤ BMI < 40	BMI ≥ 40	Total
<50 years	357 (53.5%)	188 (28.2%)	110 (16.5%)	12 (1.8%)	667 (100%)
≥50 years	158 (51.0%)	90 (29.0%)	59 (19.0%)	3 (1.0%%)	310 (100%)
Unknown	1 (100%)	0	0	0	1 (100%)
Total	516 (52.8%)	278 (28.4%)	169 (17.3%)	15 (1.5%)	978 (100%)

BMI, body mass index.

Table 3 Age at diagnosis of BC and type of institution

	Age at time of study	Age at diagnosis	Reported year of diagnosis	Type of institution
Case number				
1	45	42	2006	CSRF
2	47	45	2007	CSRF
3	50	28	1987*	CSRF
4	50	49	2008	Specialised
5	51	46	2004	CSRF
6	52	35	1992*	Specialised
7	54	45	2000	Specialised
8	58	51	2002	CSRF
9	58	56	2007	Specialised
10	59	49	1999	CSRF
11	59	51	2001	CSRF
12	62	?	?*	CSRF
13	63	63	2009	CSRF
14	71	61	1999	CSRF
Average	55.6	47.8		

CSRF, community-style residential facilities; Specialised, specialised medical institution.
*Patients not included in the calculation of standardised incidence ratio.

- *Comparison between patients with and without breast cancer*

Table 4 compares the characteristics of the 14 women with BC to the rest of the study population. Obesity was twice as frequent in women with BC as compared to those without (42.9% vs. 18.7%, $P = 0.0196$). There was no difference in terms of physical exercise during the previous 12 months between those with BC (42.9%) and those without (46.4%, $P = 0.62$). None of the women who had BC were in a relationship, and five were employed. Four were living in specialised institutions, and 10 were living in community-style residential facilities,

without a significant difference ($P = 0.11$) between those with BC and those without.

Breast cancer screening

Among the 978 women with ID in the study, 408 (41.7%) declared that they had undergone at least one mammogram, while 501 had never had any and no data were obtained for 69. Among the 310 women with ID aged over 50, and thus eligible for organised screening, 238 (76.8%) had already had at least one mammogram, 54 had not had any exam and 18 did not know. Among the 667 women aged less than 50 years, 170 (25.5%) had had at least one mammogram (Table 5).

Table 6 compares the characteristics of women with ID aged ≥50 who had already had at least one mammogram, with the rest of the study population. Among the 238 women who had already undergone a mammogram, 199 had had it within the previous 2 years, yielding an adherence rate of 64.2% (199/310) to the recommended national screening schedule.

Women with ID aged ≥50 years who were employed participated significantly more often in BC screening than those who did not work (90.7% vs. 73.8%, $P = 0.0127$); findings were similar among those aged <50 years (33.6% vs. 23.1%, $P = 0.0281$). Furthermore, women aged ≥50 and living in community-style residential facilities participated significantly more often in BC screening than those living in specialised institutions (85.9% vs. 68.3%, $P = 0.001$), and again, the same was valid for women aged <50 years (30.8% vs. 19.8%, $P = 0.0016$). There was no difference in screening participation according to obesity (78.7% vs. 76.3%, obese vs. non-obese, $P = NS$). Similarly, there was no difference in screening participation in women aged ≥50 according to whether or not they had a legal guardian (92% vs. 75.7%, $P = NS$).

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Table 4 Characteristics of the study population with vs. without cancer at the time of the survey

		N	Breast cancer				P value
		Total	No		Yes		
			N	%	N	%	
N		978	964	—	14	—	
Age category							P = 0.0064
	15–19	2	2	0.2%	0	—	
	20–24	66	66	6.8%	0	—	
	25–29	99	99	10.3%	0	—	
	30–34	96	96	10.0%	0	—	
	35–39	115	115	11.9%	0	—	
	40–44	145	145	15.0%	0	—	
	45–49	144	142	14.7%	2	14.3%	
	50–54	135	130	13.5%	5	35.7%	
	55–59	105	101	10.5%	4	28.6%	
	60–64	49	47	4.9%	2	14.3%	
	65–69	13	13	1.3%	0	—	
	70–74	7	6	0.6%	1	7.1%	
	75 and over	1	1	0.1%	0	—	
	Unknown	1	1	0.1%	0	—	
Average age		42.8	42.6	—	47.67	—	
Age category							P < 0.0001
	15–49 years	667	665	69.1%	2	14.3%	
	50 and over	310	298	30.9%	12	85.7%	
Body mass index							P = 0.072
	<25	516	510	52.9%	6	42.9%	
	25 to 29	276	274	28.4%	2	14.3%	
	30 to 39	170	165	17.1%	5	35.7%	
	≥40	16	15	1.6%	1	7.1%	
Obesity							P = 0.0196
	No (BMI < 30)	792	784	81.3%	8	57.1%	
	Yes (BMI ≥ 30)	186	180	18.7%	6	42.9%	
Type of institution							P = 0.11
	CSRF	494	484	50.2%	10	71.4%	
	Specialised	484	480	49.8%	4	28.6%	
Employed							P = 0.18
	No	768	759	78.7%	9	64.3%	
	Yes	209	204	21.2%	5	35.7%	
Relationship							P = 0.87
	No	959	945	98.0%	14	100.0%	
	Yes	18	18	1.9%	0	0.0%	

Discussion

Frequency

This is the first study to describe frequency of BC and BC screening, among a representative national sample of women with ID living in institutions. With 11 incident cases of BC (and possibly 12) observed between 1999 and 2009, for an expected 12.8 cases, the incidence of BC among women with ID living in institutions is similar to that of the general population (SIR 0.857, 95%CI 0.42–1.53). A survey performed in Finland among 1083 women with ID found a similar SIR to ours, at 0.9 (0.6–1.3), with 23 observed BC cases for 26 expected (Patja *et al.* 2001). Another

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Table 5 Number and percentage of women with intellectual disabilities aged 50 years and over who had already undergone at least one mammogram

Age	Yes		No		Don't know		Total	
	n	%	n	%	n	%	n	%
<50 years	170	25.5%	447	67.0%	50	7.5%	667	100%
≥50 years	238	76.8%	54	17.4%	18	5.8%	310	100%
Unknown	0	0	0	0	1	100%	1	100%
Total	408	41.7%	501	51.2%	69	7.1%	978	100%

Table 6 Characteristics of women with intellectual disabilities aged 50 years and over according to whether or not they had undergone at least one mammogram

		N	Mammogram				P value
		Total	No		Yes		
			N	%	N	%	
N		310	72	23.2%	238	76.8%	
Down's syndrome		35	10	28.6%	25	71.4%	
Autism		25	9	36.0%	16	64.0%	
Age category							
	50–54	135	30	22.2%	105	77.8%	
	55–59	105	21	20.0%	84	80.0%	
	60–64	49	15	30.6%	34	69.4%	
	65–69	13	2	15.4%	11	84.6%	
	70–74	7	3	42.9%	4	57.1%	
	75 and over	1	1	100%	0	0%	
Obesity							P = NS
	No (BMI < 30)	249	59	23.7%	190	76.3%	
	Yes (BMI ≥ 30)	61	13	21.3%	48	78.7%	
Employed							P = 0.0127
	No	256	67	26.2%	189	73.8%	
	Yes	54	5	9.3%	49	90.7%	
Type of institution							P = 0.001
	CSRF	149	21	14.1%	128	85.9%	
	Specialised	161	51	31.7%	110	68.3%	
Date of last mammogram							
	Mammogram = Yes				238	100.0%	
	Within previous 2 years				199	83.6%	
	Within previous 3 years				16	6.7%	
	More than 3 years ago				7	3.0%	
	Don't know				16	6.7%	

NS, non-significant; BMI, body mass index; CSRF, Community-style residential facilities; Specialised, specialised medical institution.

study in Australia among 3919 women with ID found a SIR of 0.69 (0.43–1.06), with 21 observed BC for 30 expected cases (Sullivan *et al.* 2004). According to these authors, the observed rate of BC was probably underestimated because of lower participation of women with ID in screening. A study from an

institution in the Netherlands reported a SIR of 1.24 (0.52–2.28) among 463 women with ID, with eight observed cancers for 6.43 expected cases (Evenhuis *et al.* 1996). Similarly, a study in a French hospital including 484 invasive BC cases reported that 11 (2.23%) were women with ID, which approximately corresponds to the estimated frequency of ID in the general population (Satgé *et al.* 2014). All these studies concern somewhat different populations to that included in our study, as regards the level of ID and the living conditions. However, all the studies, including our own, suggest that the risk of BC among women with ID is similar to that of women in the general population. This risk could even be somewhat higher, since the incidence of BC increases with age, and the life expectancy of women with ID has increased since the publication of the studies cited above and, indeed, continues to progress (Tuffrey-Wijne *et al.* 2009). In our study, BC was the most common type of cancer observed, underlining the necessity to accord particular attention to this disease in this population.

Age at diagnosis

The average age of patients at the time of diagnosis in our study was 47.8 years, which is unusually low compared to the average age at diagnosis for BC in France, which was 63 years of age for the general population during the study period (Binder-Foucard *et al.* 2014). This discrepancy can likely be explained by the difference in age distribution of the 978 women with ID included in our study as compared to the general population of women in France. Indeed, our population of women with ID was particularly young, with an under-representation of the older age groups, and an over-representation of the younger age groups as compared to the general population. The calculation of the expected number of BC adjusted by age class did not show any significant difference compared to the general population. Furthermore, our data suggest that the cumulative risk of experiencing BC before the age of 50 years among women with ID living in institutions (2.03%, 0.4%–3.66%) is not significantly different from that of the general population (2.4%, 1%–3.78%). Therefore, our study shows that the younger age observed at diagnosis is due to the fact that, overall, the population of our study is younger. A previous French

study reported that among 11 women with ID treated in a single hospital, the average age at diagnosis was 55.6 years, vs. 62.4 in a control group, and also found that women with ID had more advanced cancer stage at diagnosis (Satgé *et al.* 2014). Similarly, an Australian study of 19 patients reported an average age of 49 years at diagnosis (range, 29–86) for the period 1982–2000 (Sullivan *et al.* 2003). Conversely, a report from the Netherlands found that among eight patients with ID, average age at BC diagnosis was 60 years (Evenhuis *et al.* 1996). Further studies are warranted to investigate the average age at onset of BC in this specific population, as this information is fundamental to planning appropriate surveillance and screening.

Risk factors

In addition to genetic predispositions, major risk factors for BC include early puberty, late menopause, few or no pregnancies, never having breastfed, obesity, sedentary lifestyle, alcohol consumption, smoking and hormone replacement therapy (Howell *et al.* 2014). In our study, the questionnaire did not include any questions pertaining to family history of BC, or use of oral contraceptive pills. Women with ID generally consume little, if any alcohol and tobacco, and do not take hormone replacement therapy. However, this group as a whole is theoretically at increased risk of BC since pregnancy and breastfeeding are rare, and obesity and sedentarity are more common than in the general population (McGuire *et al.* 2007). To the best of our knowledge, ours is the first study to report on obesity as a risk factor for BC among women with ID living in institutions. Among the 14 women with BC, six were obese (42.9%), and this rate was significantly higher than the population without BC, where 18.7% had a BMI ≥ 30. In the general French population, there were an estimated 6.5 million obese people in 2009, corresponding to 14.5% of the French population (France Key Facts 2014).

It is well established that the rate of obesity is high among women with ID (Rimmer & Yamaki 2006; McGuire *et al.* 2007). This can be explained by certain predisposing genetic conditions, such as Down syndrome, Prader–Willi syndrome or others (Fox & Rotatori 1982; Rubin *et al.* 1998). Environmental and lifestyle factors constitute another

major contributor, such as a rich diet and a low level of physical exercise (Temple & Walkley 2003; McGuire *et al.* 2007). Accordingly, obesity is more frequent among people with ID who live at home with their family as compared to those who live in an institution (Prasher 1995; Rubin *et al.* 1998). Obesity, particularly after the menopause, increases the relative risk of BC in line with the degree of excess weight, with a relative risk of 1 for a BMI < 25, increasing to 1.21 for overweight, and 1.29 for obesity (Eliassen *et al.* 2006). A further step towards understanding the role of obesity could be to evaluate the frequency of BC and age at diagnosis in women with ID living at home with their family, since obesity is more frequent in this subgroup of women with ID. The very small number of women living maritally in this institutional population underlines the numeric magnitude of the risk linked to the lack of pregnancy or breastfeeding.

Screening

Among the 310 women aged ≥50 in our study population, who were eligible for the national screening programme, 76.8% had already had at least one mammogram, and 199 had had it within the previous two years, which is in line with the recommendations for the screening interval. These findings suggest that women with ID living in institutions actively participate in organised screening (64.2%). The overall participate rate for women aged between 50 and 74 years in the general population was estimated to be 62% in 2009–2010, composed of 52% through organised screening, and 10% through individual screening.

The participation rate observed in our study is higher compared to two other studies from the French region of Provence-Alpes-Cote d'Azur. One of these studies reported that only 2.2% of general practitioners oriented women with ID towards organised screening (Verger *et al.* 2005). The second study reported that 52% of institutions for women with mental or physical impairments ensured active participation of all eligible residents in BC screening (Couepel *et al.* 2011). Similarly, studies from the USA (Havercamp *et al.* 2004; Iacono & Sutherland 2006; Parish & Saville 2006), Australia (Davies & Duff 2001), the United Kingdom (Osborn *et al.* 2012) and Canada (Cobigo *et al.* 2013) have all reported lower

participation among women with ID in institutions as compared to the general population, with rates ranging from 19.4% to 87% for women with ID living in institutions, in their family environment or in the community. Another study from the United Kingdom found a participation rate similar to that of the general population (77% vs. 75%) (Biswas *et al.* 2005). In our study, the rate of participation in screening was slightly higher than that of the general French female population for the same period (64.2% vs. 62%). Son *et al.* (2013) previously reported that women with ID declared that they underwent more mammograms than was actually the case. However, in our study, the interviews were assisted by a professional from the residential institution, which limits the potential for over-estimation of the number of screening exams reported. Although similar to that of the French general population, the participation rate is far from being optimal. It suggests that nearly 36% of women living in an institutional setting, and who have difficulties communicating about their unease and their disease, do not currently receive the health benefits from BC screening as their peers with greater levels of ability.

Eight cancers (57.2%) were diagnosed in women aged <50 years in our study, that is before the age at which they would have become eligible for screening. This suggests that the diagnosis was based on clinical signs. However, given the high number of women aged less than 50 who had already undergone at least one mammogram in this study (25.5% in all, of which 37% aged 40–44, and 48% aged 45–49 years), we cannot draw any conclusions about the mode of discovery of these cancers (palpation or individual screening).

Special populations

Women with ID aged 50 years and older who were living in specialised institutions participated significantly less often in organised screening than women with ID who were living in community-style residential facilities (68.3% vs. 85.9%, $P = 0.001$), even though this rate of participation remained above the national average. This difference could be explained by the fact that such specialised institutions are generally for patients with more severe disabilities, often with comorbidities and more handicaps than those living in community-style residential facilities, thus making it more difficult to perform

mammograms. It is also possible that due to negative attitudes and other barriers to health care, medical professionals or support staff may feel that BC screening among women with severe disability is not warranted because they would not be offered treatment if diagnosed with this condition (Kiani *et al.* 2014). This suggests that a special effort should be made to target women living in specialised medical institutions, possibly by reasonable adjustment to traditional methods, such as using other diagnostic methods to screen for BC (ultrasound, palpation). More than 90% of women with ID who were employed participated in screening after the age of 50 in our study, and the rate of participation was significantly higher than those who were not employed (90.7% vs. 73.8%, $P = 0.0127$). This may be linked to better medical follow-up, in particular through work-related medical exams. Indeed, women with ID who were employed were more independent, had fewer disabilities and, therefore, were better able to participate in screening. This underlines the importance of targeting women who are not employed for screening programmes.

Our study was performed among women with ID living in institutions, and accordingly, in this group, there was an over-representation of women with severe and moderate disabilities as compared to milder handicaps. In this regard, our study does not inform about women with ID living at home with their family, or living in the community. This study does provide, for the first time, a quantitative estimate of the frequency of BC among women with ID living in institutions in France and shows that the risk is similar to that of women in the general population. Our results also represent the first documented confirmation of the importance of obesity, nulliparity and absence of breastfeeding as risk factors in this population.

The frequency of BC largely justifies the implementation of screening programmes among women with ID, as in the general population, as well as the use of alternative methods such as ultrasound or palpation when mammography is not possible (Willis *et al.* 2008). For these women, screening is all the more important and useful because they have little knowledge about the disease (Gillings-Taylor 2008; Poynor 2003; Truesdale-Kennedy *et al.* 2011; Wilkinson *et al.* 2011). If they notice a lump on their breast, they are not as likely to consult a doctor about

it as a person without disability. Consequently, many tumours among women with ID are discovered at a more advanced stage as compared to women in the general population (Tuffrey-Wijne 1997; Satgé *et al.* 2014). In order to enhance prevention of BC, as with other types of cancer, it is important to reduce obesity and encourage regular exercise (Maïano *et al.* 2014).

Study limitations

This study is based on self-reported data. All the cases of cancer reported here were not verified and validated. We did not collect data about the tumour status (histology, stage and grade in particular), or the management of women who declared having BC in this study. The malignant nature of the tumours was not confirmed. There may have been *in situ* cancers that would not be included in the estimation of incident cases provided by cancer registries, which would influence the calculation of the SIR. Conversely, it is also possible that some cancers went undeclared, because the person responding on behalf of the woman with ID was unaware of it, although this is unlikely. Furthermore, women with ID living in institutions who had had BC and had died were obviously not included in this survey. We also do not have any detail of the proportion of organised screening and the proportion of individual screening among the women with ID who had undergone a mammogram. The strong points of this study include its wide coverage of institutions across the whole country, the different types of institutions included and the random selection of participants, strongly supporting the claim that the sample in this study was representative.

Conclusion

In this sample of 978 women with ID living in institutions in France, we show that the rate of BC is similar to that in the general population. The frequency of BC is likely to increase with the constant progression in life expectancy among this population. In our study, contrary to previous reports, we did not find that BC occurred any earlier than in the general population. Our results confirm the importance of established risk factors such as obesity or nulliparity. The rate of participation in screening of women with ID living in institutions was found to be slightly

higher in this study than that of the general population. This participation should be enhanced in a group of women who do not easily communicate their symptoms and who are at risk of delayed diagnosis. The participation rate could particularly be enhanced among residents of specialised medical institutions for persons with severe disabilities, and among women who are not employed. The confirmation of the importance of obesity as a risk factor provides an avenue for preventive measures. Finally, further studies are warranted to improve our knowledge of the characteristics of cancer at the time of diagnosis in this particular population.

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References

Anampa J., Makower D. & Sparano J. A. (2015) Progress in adjuvant chemotherapy for breast cancer: an overview. *BMC Medicine* **13**, 195.

Binder-Foucard F., Bossard N., Delafosse P., Belot A., Woronoff A. S. & Remontet L. (2014) French network of cancer registries (Francim). Cancer incidence and mortality in France over the 1980–2012 period: solid tumors. *Revue d'Épidémiologie et de Santé Publique* **62**, 95–108.

Biswas M., Whalley H., Foster J., Friedman E. & Deacon R. (2005) Women with learning disability and uptake of screening: audit of screening uptake before and after one to one counselling. *Journal of Public Health (Oxford, England)* **27**, 344–7.

Bourgarel S., Trétarre B., Satgé D. & Stoebner-Delbarre A. (2015) Le cancer colorectal et son dépistage chez les personnes déficientes intellectuelles vivant en institution en France. *Oncologie*. DOI: 10.1007/s10269-015-2532-1.

Bouvier G. (2011) Le volet institution de l'enquête Handicap-Santé: Présentation, calcul des poids. Document de travail INSEE F1102 pp 29

Brownstein M. H., Wolf M. & Bikowski J. B. (1978) Cowden's disease: a cutaneous marker of breast cancer. *Cancer* **41**, 2393–8.

Cobigo V., Ouellette-Kuntz H., Balogh R., Leung F., Lin E. & Lunsky Y. (2013) Are cervical and breast cancer screening programmes equitable? The case of women with intellectual and developmental disabilities. *Journal of Intellectual Disability Research* **57**, 478–88.

Couepel L., Bourgarel S. & Piteau-Delord M. (2011) Dépistage du cancer chez les personnes handicapées: pratiques et difficultés en établissement médicosocial. *Prat et Org soins* **42**, 245–53.

Cowppli-Bony A., Uhry Z., Remontet L., Guizard A. V., Voirin N., Monnereau A. et al. (2016) Survie des personnes atteintes de cancer en France métropolitaine, 1989–2013. Partie 1 – Tumeurs solides. Saint-Maurice (Fra): *Institut de Veille Sanitaire*.

Davies N. & Duff M. (2001) Breast cancer screening for older women with intellectual disability living in community group homes. *Journal of Intellectual Disability Research* **45**, 253–7.

Eliassen A. H., Colditz G. A., Rosner B., Willett W. C. & Hankinson S. E. (2006) Adult weight change and risk of postmenopausal breast cancer. *JAMA* **296**, 193–201.

Evenhuis H. M., Oostindïer M. J., Steffelaar J. W. & Coebergh J. W. (1996) Incidentie van kanker bij mensen met een verstandelijke handicap; mogelijk verhoogd risico op slokdarmkanker. *Nederlands Tijdschrift voor Geneeskunde* **140**, 2083–7.

Farach A., Farach L. S. & Paulino A. C. (2013) Therapeutic challenges in treating patients with fragile X syndrome and neoplasia. *Pediatric Blood & Cancer* **60**, E153–6.

Foca F., Mancini S., Bucchi L., Puliti D., Zappa M., Naldoni C. et al. (2013) Decreasing incidence of late-stage breast cancer after the introduction of organised mammography screening in Italy. *Cancer* **119**, 2022–8.

Fox R. & Rotatori A. F. (1982) Prevalence of obesity among mentally retarded adults. *American Journal of Mental Deficiency* **87**, 228–30.

France Key Facts (2014) Obesity and the economics of prevention: fit not fat – http://www.oecd.org/els/health-systems/obesityandtheeconomicsofpreventionfitnotfat-francekeyfacts.htm.

Gillings-Taylor S. (2008) Why the difference? Advice on breast examination given to carers of women who have learning disability and to women who do not. *Journal of Learning Disabilities* **8**, 175–89.

Haute Autorité de Santé (2011) http://www.hassante.fr/portail/upload/docs/application/pdf/2012-07/synthese_et_recommandations_participation_depistage_cancer_du_sein.pdf.

Havercamp S. M., Scandlin D. & Roth M. (2004) Health disparities among adults with developmental disabilities, adults with other disabilities, and adults not reporting disability in North Carolina. *Public Health Reports* **119**, 418–26.

Hirtzlin I., Barré S. & Rudnichi A. (2012) Dépistage individuel du cancer du sein des femmes de 50 à 74 ans en France en 2009. *BEH* **35-36-37**, 410–412.

Howell A., Anderson A. S., Clarke R. B., Duffy S. W., Evans D. G., Garcia-Closas M. *et al.* (2014) Risk determination and prevention of breast cancer. *Breast Cancer Research* **16**, 446.

Iacono T. & Sutherland G. (2006) Health screening and developmental disabilities. *Journal of Policy and Practice in Intellectual Disabilities* **3**, 155–64.

Jancar M. P. & Jancar J. (1977) Cancer and mental retardation. *Bristol Med-Chir* **92**, 3–7.

Kiani R., Tyrer F., Shaikh A. & Satgé D. (2010) Cancer deaths in people with moderate to profound intellectual disabilities: the Leicestershire experience 1993–2006. *International Journal of Child Health and Human Development* **3**, 215–21.

Kiani R., Vahabzadeh A., Hepplewhite E. A., Abbas M., Finnamore T., Bhaumik S. *et al.* (2014) Overcoming challenges in diagnosing and treating cancers in people with intellectual disability: a case analysis. *Tizard Learning Disability Review* **19**, 51–8.

Maïano C., Normand C. L., Aimé A. & Bégarie J. (2014) Lifestyle interventions targeting changes in body weight and composition among youth with an intellectual disability: a systematic review. *Research in Developmental Disabilities* **35**, 1914–26.

McGuire B. E., Daly P. & Smyth F. (2007) Lifestyle and health behaviours of adults with an intellectual disability. *Journal of Intellectual Disability Research* **51**, 497–510.

Osborn D. P., Horsfall L., Hassiotis A., Petersen I., Walters K. & Nazareth I. (2012) Access to cancer screening in people with learning disabilities in the UK: cohort study in the health improvement network, a primary care research database. *PLoS One* **7**, e43841.

Parish S. L. & Saville A. W. (2006) Women with cognitive limitations living in the community: evidence of disability-based disparities in health care. *Mental Retardation* **44**, 249–59.

Patja K., Eero P. & Iivanainen M. (2001) Cancer incidence among people with intellectual disability. *Journal of Intellectual Disability Research* **45**, 300–7.

Poynor L. (2003) Being breast aware. *Learning Disability* **6**, 10–4.

Prasher V. P. (1995) Overweight and obesity amongst Down's syndrome adults. *Journal of Intellectual Disability Research* **39**, 437–41.

Rimmer J. H. & Yamaki K. (2006) Obesity and intellectual disability. *Mental Retardation and Developmental Disabilities Research Reviews* **12**, 22–7.

Rogel A., Lastier D. & Saline E. (2012) Évaluation du Programme national de dépistage organisé du cancer du sein en France: période 2004–2009. *BEH* **35-36-37**, 399–404.

Rubin S. S., Rimmer J. H., Chicoine B., Braddock D. & McGuire D. E. (1998) Overweight prevalence in persons with Down syndrome. *Mental Retardation* **36**, 175–81.

Sahlin P., Windh P., Lauritzen C., Emanuelsson M., Grönberg H. & Stenman G. (2007) Women with Saethre–Chotzen syndrome are at increased risk of breast cancer. *Genes, Chromosomes & Cancer* **46**, 656–60.

Satgé D., Sasco A. J., Azema B. & Culine S. (2007) Cancers in persons with intellectual deficiency: current data. In: *Mental Retardation Research Focus* (ed. M. V. Charleton), pp. 47–84. Nova Sciences Publisher, New York.

Satgé D., Sasco A. J., Pujol H. & Rethoré M. O. (2001) Les tumeurs mammaires chez les femmes trisomiques 21. *Bulletin de l'Académie Nationale de Médecine* **185**, 1239–54.

Satgé D., Sauleau E. A., Jacot W., Raffi F., Azéma B., Bouyat J. C. *et al.* (2014) Age and stage at diagnosis: a hospital series of 11 women with intellectual disability and breast carcinoma. *BMC Cancer* **14**, 150.

Sharif S., Moran A., Huson S. M., Iddenden R., Shenton A., Howard E. *et al.* (2007) Women with neurofibromatosis 1 are at a moderately increased risk of developing breast cancer and should be considered for early screening. *Journal of Medical Genetics* **44**, 481–4.

Son E., Parish S. L., Swaine J. G. & Luken K. (2013) Accuracy of self-reported cervical and breast cancer screening by women with intellectual disability. *American Journal on Intellectual and Developmental Disabilities* **118**, 327–36.

Sullivan S. G., Glasson E. J., Hussain R., Petterson B. A., Slack-Smith L. M., Montgomery P. D. *et al.* (2003) Breast cancer and the uptake of mammography screening services by women with intellectual disabilities. *Preventive Medicine* **37**, 507–12.

Sullivan S. G., Hussain R., Threlfall T. & Bittles A. H. (2004) The incidence of cancer in people with ID. *Cancer Causes and Control* **15**, 1021–5.

Temple V. A. & Walkley J. W. (2003) Physical activity of adults with intellectual disability. *Journal of Intellectual & Developmental Disability* **28**, 342–52.

Truesdale-Kennedy M., Taggart L. & McIlfatrick S. (2011) Breast cancer knowledge among women with intellectual disabilities and their experiences of receiving breast mammography. *Journal of Advanced Nursing* **67**, 1294–304.

Tuffrey-Wijne I. (1997) Palliative care and learning disabilities. *Nursing Times* **5**, 50–1.

Tuffrey-Wijne I., Bernal J., Hubert J., Butler G. & Hollins S. (2009) People with learning disabilities who have cancer:

an ethnographic study. *British Journal of General Practice* **59**, 503–9.

Verger P., Aulagnier M., Souville M., Ravaud J. F., Lussault P. Y., Garnier J. P. *et al.* (2005) Women with disabilities: general practitioners and breast cancer screening. *American Journal of Preventive Medicine* **28**, 215–20.

Wilkinson J. E., Deis C. E., Bowen D. J. & Bokhour B. G. (2011) 'It's easier said than done': perspectives on mammography from women with intellectual disabilities. *Annals of Family Medicine* **9**, 142–7.

Willis D. S., Satge D. & Sullivan S. G. (2008) Breast cancer surveillance in women with learning disabilities. *International Journal on Disability and Human Development* **7**, 405–11.