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Self-injurious behavior: A comparison between Prader-Willi syndrome, Down syndrome and Autism

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Abstract

The term Self-injurious Behavior (SIB) represents behavioral characteristics that can damage body tissue. Persons with Intellectual Disability (ID) have a greater probability of developing inadequate behavior such as SIB. Literature indicates the presence of SIB in different syndromes. In the Prader-Willi Syndrome (PWS) the most frequent physical manifestation is skin-picking, which involves the arms, legs, and hands. In the Down Syndrome (DS), eyes and ears are more greatly affected by SIB. Prevalent SIB regards head-hitting and eyes. In subjects with Autistic Disturbances (AD), effects of SIB can be found more frequently in the head, hands, and legs. The more common typologies of SIB are self-biting and self-hitting.
The sample was composed of 84 persons with ID and associated syndromes (PWS, DS, AD). The SIB has been identified through administering a specific assessment instrument to caregivers.
The collected data show that self-inflicted injuries are present in all three groups. In groups with PWS and AD most common examples regard the hands, respectively 90% and 55%; subjects with DS show SIB on the mouth (48%). SIB most frequently shown in subjects with PWS are "skin-picking" (50%), in subjects with DS grinding their teeth (32%), in those with AD the prevalent SIB is "body-hand-hitting" and "body-object hitting" (41%).

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Our data show some convergences with results from literature (e.g., we confirmed that in PWS the most common manifestation of SIB is "skin-picking"), while other results are not confirmed. Differences could be explained according to more restricted or wide definitions of SIB.

Keywords: Self-Injurious Behavior, Intellectual Disability, Prader-Willi Syndrome, Down Syndrome, Autistic Disorder.

1. Introduction

The definition of Self-Injurious Behavior (SIB) is an object of debate in literature. Varying degrees of broadly-defined terms for SIB exist which determine a significant variability in data about typology and prevalence.

Many Authors have elaborated differing terms (self-harm behaviors, self-injurious behaviors, self-mutilation) for behavioral symptoms that often are similar (Palmigiano, Scannella, & Buono, 2007). Others have defined self-inflicted injury as behavior that produces immediate and cumulative physical damage to one's own body (Tate & Baroff, 1966). The SIB is often shown in diverse clinical conditions - especially when they are characterized by severe pathologies - such as epilepsy, muscle-nerve disorders, hearing diseases, communicative disabilities, stereotypical behaviors (Emerson, Kiernan, Alborz, Reeves, Mason, Swarbrick *et al.,* 2001).

The SIB often can block the process of one's personal growth and the opportunity to integrate into society subjects with these disorders; besides this, it can be the reason for the failure of rehabilitation and integration efforts in community contexts (Hill & Bruininks, 1984).

Persons with Intellectual Disability (ID) are more vulnerable to psychopathologies. In such groups of persons, the comorbidity is estimated three or four times superior respect to the general population.

During these subjects' lifespan, some can develop behavioral disturbances at different levels manifesting themselves in the form of aggressive and destructive conduct, violent fits of anger, stereotyped or self-injuring behavior (Symons & Thompson, 1997; Rojahn, Tassé, & Morin, 1998; American Psychiatric Association, 2000).

The most common forms of SIB associated with ID, as shown in literature, seem to be: self-biting, head-banging, and self-scratching. As cited above, there is little agreement about hair pulling (Arzeno Ferrao, Almeida, Bedin, Rosa, & D'Arrigo Busnello, 2006), onicofagy (Wells, Haines, Williams, & Brain, 1999), and sometimes skin-picking (Keuthen, Wilhelm, Deckersbach, Engelhard, Forker, Baer *et al.,* 2001; Singareddy, Moin, Spurlock, Merritt-Davis, & Uhde, 2003), considered often as phenomena explained in a spectrum of obsessive-compulsive behaviors, or as deficit of impulse control.

In DSM-IV-TR (American Psychiatric Association, 2000) the term self-inflicted injury can be adopted in disturbances of stereotyped movements described as repeated motor-nerve behavior apparently intentional and afinalistic which for themselves can be specified as self-injury behavior if such behavior can cause physical damage which requires specific treatment, or could cause physical injury if protective measures are not taken.

The 10th Revision of The Classification of Mental and Behavioural Disorders: Clinical Descriptions and Diagnostic Guidelines (World Health Organization, 1992) inserts self-inflected injury as disturbance caused by

Stereotyped Movements", i.e., movements which are "voluntary, repetitive, stereotyped, non functional", which can be subdivided into "self-infliction" and "non self-infliction. The first group includes repeated head-hitting, slapping, eye-hitting, hand, lip, body- biting.

According to DSM-IV-TR there doesn't exist a "typical" age or modality for the initial disturbance of stereotyped movement, of which self-infliction is integral part. Besides this, reference is made to the fact that localization of this behavior is modified through the years (American Psychiatric Association, 2000).

Various studies have shown that there is an initial phase of SIB during childhood (Oliver, Murphy, & Corbett, 1987; Keuthen *et al.,* 2001).

Other studies (Rojahn, 1986; Kahng, Iwata, & Lewin, 2002) have shown that more frequent forms of self-infliction are: head and body-hitting, self-biting, skin-picking, scratching the body, self-pinching.

Additional studies have tried to reveal the locations that are most often object of self-infliction. It appears that such zones are distributed irregularly on the human body and more often on the head and hands. In the specific case of the head, the area most frequently hit seems to be the face, and on the back of the hand. The other zones are distributed on the back and lower arms, as well as the back of the legs (Symons & Thompson, 1997).

Self-infliction can be a psychopathological characteristic frequently associated with diverse syndromes with multiple etiology.

Among the syndromes in which the presence of SIB has been indicated we will discuss the Prader-Willi Syndrome the Down Syndrome, and Autism.

1.1 Prader-Willi Syndrome

The Prader-Willi Syndrome (PWS) is caused by the deletion in the 15q11-q13 region of chromosome 15 of paternal derivation.

The distinctive characteristics of PWS are hypotony, obesity, hypo-gonadism, intellectual disability, mood instability, temper tantrums (Clarke, Boer, Chung, Sturmey, & Webb, 1996; Plantin, Milochau, Broussine, & Blondin, 1997; Symons, Butler, Sanders, Feurer, & Thompson, 1999; Campeotto, Naudin, Viot, & Dupont, 2001; Whitman, Myers, Carrel, & Allen, 2002). In literature it has been reported that subjects with PWS show severe self-infliction with a prevalence that varies from 70% to 90% (Symons *et al.,* 1999; Wigren & Heimann, 2001; Buono, Palmigiano, & Scannella, 2005).

Symons *et al.*, (1999) have revealed that the most common form of self-inflection is skin-picking that is manifested in 69% of young people with PWS and 81% of adults. The head (42%), legs (40%) and arms (39%) are the body areas most often hit, and in particular the rear areas. The Authors do not seem to have noted differences correlated to gender in relation to the number of parts hit. Adults frequently hurt themselves in more than one area of the body.

In a sample of subjects with PWS and intellectual disability, Buono *et al.*, (2005) have reported that 73% initiated self-injurious behavior.

Wigren and Hansen (2003) have outlined that there exists a co-existence between compulsive and self-injurious behavior, more frequently with skin-picking and hair-pulling. Other authors have also discussed this concordance of factors (Stein, Keating, Zar, & Hollander, 1994). The authors identify skin-picking in a specific compulsive dimension.

Dimitropoulos, Feurer, Butler and Thompson (2001) have shown that in a sample of 105 subjects with PWS, a third put into act SIB of a skin-picking type, coherent with data originating from literature; while the relationship between chronological age and skin picking is not confirmed.

Subjects with PWS derived from uni-parental disomy show diverse behavioral dysfunctions among which is SIB manifested through some habits, as skin-picking and nail biting. People with PWS derived form deletion are predisposed to initiate SIB of the type: skin-picking, nail-biting, hair-pulling. According to the animal model of compulsive SIB, serotoninergic and dopaminergic mechanisms assume an important role (Dykens, Cassidy, & King, 1999).

1.2 Down Syndrome

Down syndrome is the most common genetic cause of intellectual disability, occurring in approximately 1 in 700 live births (Centres for Disease Control and Prevention, 2006).

Ninety-eight percent of cases of Down syndrome are caused by an extra copy of chromosome 21 (Trisomy 21) (Hassold & Sherman, 2002).

The DS is one of the most frequent causes of intellectual disability. It can be recognized at birth for its phenotypic characteristic to which can be added delay in psychic-motor nerve growth, Intellectual Disability at a different level, cardiopathy, epilepsy, breathing pathologies, behavioral problems, hard-headedness, emotive disturbances, dementia (Korenberg, Chen, Schipper, Sun, Gonsky, Gerwehr *et al.,* 1994).

As for phenotypic aspects, subjects with DS can be characterized by their naturally warm and a generally peaceful nature and ability to mimic others (Menolascino, 1965; Gibbs & Thorpe, 1983). But sometimes behavioral problems such as aggressiveness, and highly pronounced hyperactivity may be present (Menolascino, 1965, 1967; Gath & Gumley, 1984; Myers & Pueschel, 1991; Cuskelly & Dadds, 1992).

In literature, a prevalence of 25% comorbid psychological disturbances is forecast, even if the data varies at diverse levels. Behavioral disturbances are diagnosed in over 10% of children. The same percentage is revealed for attention/hyperactivity deficit, mood disorders, association with autism and infantile psychosis (Menolascino, 1965, 1967; Gath & Gumley, 1986; Howlin, Wing, & Gould, 1996). Several authors have emphasized a difference due to gender for vulnerability to psychopathological problems (Menolascino, 1965; Gath & Gumley, 1986).

Literature presents examples of cases that show multiple SIB that affect various regions of the body; i.e. hitting the eyes, generating contusions and lacerations hitting the head, body, face, ears with the hands, and also hitting the head toward object. The body areas more greatly affected by the SIB syndrome seem to be: lip/mouth, hands, head, and cheek.

Mazaleski, Iwata, Vollmer, Zarcone and Smith (1993) have determined the body areas affected by SIB to be hands and face.

Fisher, Bowman, Thompson, Contrucci, Burd and Alon (1998), in a study conducted on a boy with Down Syndrome, profound intellectual disability and severe SIB, have shown that the area affected was around the ears. The resulting SIB was repetitively and continually hitting the ears resulting in contusions and lacerations, and was produced once the subject had woken up.

Hagopian, Paclawskyj and Kuhn (2005) have examined the SIB behavior of a person with SD and profound intellectual disability. The areas affected were the eyes, with a definite action of eye poking. The SIB produced over time a separation of the retina.

Määttä, Tervo-Määttä, Taanila, Kaski and Iivanainen (2006) demonstrate the presence of SIB in 3% of the DS group under analysis. The SIB was associated with a clinical context of DS and severe intellectual disability. The body areas most affected were the head and ears. The resulting action was to hit and twist the ears with the hands. The resulting damage was severe. The authors have discussed the significant difficulties in treating severe forms of SIB.

Other studies have demonstrated the presence of behavioral problems such as SIB and precocious dementia in subjects with SD (Hirayama, Kobayashi, Fujita, & Fujino, 2004).

1.3 Autism

Autism Disorders (AD) is characterized by a significant qualitative and quantitative mixture of deficits. In the majority of cases this is associated with intellectual disability at a different level (American Psychiatric Association, 2000; McCracken, McGough, Shah, Cronin, Hong, Aman, *et al.*, 2002).

Various studies (Lewis & Bodfish, 1998) have demonstrated that in AD the prevalence of SIB varies from 30% to 69% (Schroeder, Schroeder, Smith, & Dalldorf, 1978; Bodfish, Symons, Parker, & Lewis, 2000).

Some studies have also demonstrated that in subjects with DA the areas most affected are the head, the hands and legs. The SIB behavior most represented is hitting oneself and self-biting (Symons, Hoch, Dahl, & McComas, 2003; Moore, Fisher, & Pennington, 2004). In a sample of 102 persons with AD, 49% of participants showed SIB behavior (Ballaban-Gil, Rapin, Tuchman, & Shinnar, 1996).

According to Baghdadli, Pascal, Grisi and Aussilloux (2003), persons with AD may develop SIB in severe forms. The factors that favor the

growth of SIB have been identified as lacking ability for autonomous action and managing daily life, besides the severity of the autism. Retarded language development and difficulty in interpersonal relationships are not considered risk factors.

Moreover, Murphy, Hall, Oliver, and Kissi-Debra, (1999) and Bodfish *et al.* (2000) have found that young age and a severe level of intellectual disability are not to be considered risk factors in developing SIB in association with DA.

Canitano (2006) conducted a study on a sample of 11 people with AD and a moderate or severe ID. All subjects presented SIB. The typologies more commonly present were head-hitting and hand-biting.

2. Aim of the study

Our study was conducted with the aim of analyze SIB in a sample constituted of subjects with differing clinical syndromes associated with ID, to reveal similarities and differences among syndromes, inherent in body localization and typology of SIB.

3. Sample

The sample was composed of 84 persons with ID (10 PWS, 25 DS, 49 AD)) selected among a wider sample constituted of 1040 persons with ID. The percentage of persons with SIB within each syndrome was 55.5% in PWS group, 45.4% in DS group, 70% in AD group.

The chronological age of the participants varied from one to 47 years of age. The PWS group consisted of 6 males and 4 females, the average age is was 14.3, SD = 7.4. The DS group was composed of 16 males and 9 females, with average age of 15.3 years, SD = 10.6, while in the autism group the males were 29, and females 20, with an average age of 13.1 years, SD = 8.5.

All subjects presented intellectual disabilities of various grades of intensity according to DSM-IV TR criteria (American Psychiatric Association 2000).

In the PWS group, 70% of subjects show mild ID, and 30% moderate. Subjects with DS are distributed in a more differentiated manner: 20% present mild IDs, 28% moderate, 44% severe, 8% profound. In the AD group, 18% had mild ID, 14% moderate, 44% severe, 24% profound.

4. Methodology

The SIB have been identified and assessed through application of a Self-Injurious Behavior Schedule (SRCA) administered, by specifically quali-

fied psychologists, to families and counselors of the observed persons, who were present in the rehabilitation wards and receive diagnostic services at the IRCCS Oasi Maria SS, Troina, Italy.

The diagnostic instrument has been previously devised and validated with the aim of revealing the presence of SIB; besides this, it is capable of discriminating between diverse topographical areas and can identify the frequency of emission and intensity of SIB. The subject's age at the moment of first SIB outbreak was also recorded.

The psychometric properties of the SRCA were assessed in a sample of 40 people with ID (Buono, Palmigiano, Scannella, & Di Nuovo, 2006). Inter-rater reliability was computed applying the schedule on single individuals on the part of two counselors.

The percentage of concordance between the two counselors resulted very high (equal to 92.33%); Cohen's *k* was .60. As regards the concurrent validation, the schedule was compared to the subscale "SIB behavior" of the AAMD test (Nihira, Foster, Shellhaas, & Leland, 1975); the percentage of concurrence between the two scales was 85%, with *k*=.65.

Informed consent has been obtained by the families of the interviewed subjects.

5. Results

The data collected in our sample demonstrate that SIB is present in all three groups taken into consideration, and that first indication of SIB (table 1) is prevalent in early infancy (<7 years). In particular in subjects with PWS, in 90% of cases SIB emerges in the above-mentioned age range; 72% for persons with DS, similar to subjects with AD (72%). In the age group 7-12 years, PWS has a SIB percentage of 10%, DS 12%, AD 10%.

Table 1 - *Distribution of sample according to the beginning age of SIB symptoms*

Beginning age	*PWS*	*DS*	*AD*
<7	90%	72%	72%
7-12	10%	12%	10%
13-18	0	4%	6%
>18	0	12%	8%
Not valuable	0	0	4%

Relative to the bodily areas affected by SIB (table 2) the distribution in the three syndromes is as following:

In the group of persons with PWS bodily areas most affected by SIB are hands (90%), arms (70%), legs (70%), mouth and head (30%), nose, cheeks, ears (20%). Persons with DS present SIB on the mouth (48%), hands (44%), head (29%), cheeks (12%), ears (8%).

In persons with DA the SIB is localized prevalently in the hands (55%), mouth (45%), head (39%), cheeks (37%), neck (16%), forehead and arms (12%).

As shown in table 2, the differences among groups in localization are statistically significant for hands and arms (both prevalent in PWS).

Tabella 2 - *Localization of SIB, differences among groups, and levels of significance to χ^2 test*

	SYNDROMES							
	PWS		*DS*		*AD*			
	N°	*%*	*N°*	*%*	*N°*	*%*	^{2}	*p*
forehead	1	10	0	0	6	12	3.29	.19
nose	2	20	0	0	4	8	4.49	.10
cheek	2	20	3	12	18	37	5.40	.06
ears	2	20	2	8	4	8	1.44	.48
eyes	0	0	0	0	1	2	0.41	.81
mouth	3	30	12	48	22	45	0.97	.61
head	3	30	5	20	19	39	2.69	.25
nape	0	0	0	0	8	16	6.31	.04
neck	1	10	0	0	3	6	2.05	.35
hands	9	90	11	44	27	55	6.16	.05
legs	7	70	0	0	10	4.9	8.59	.01
arms	7	70	0	0	6	12	27.69	<.01

As far as SIB typology is concerned (table 3), its most common manifestations in PWS are skin-picking (50%), nail picking-eating (40%), hair-pulling (30%), self-scratching and finger-object in cavities (20%)

People with DS most frequently manifest grind teeth (32%), hand hitting (28%), pick-eat nails (24%) self-biting, body-object hitting; insert finger, objects in cavities (12%).

In persons with AD the SIB most frequently manifest is hand-hitting (41%), object hitting (41%), self-biting (39%), finger-object in cavity (35%).

The significant differences among groups are shown in table 3.

Tabella 3 - *Typology of SIB, differences among groups, and levels of significance to χ^2 test*

	SYNDROMES							
	PWS		*DS*		*AD*			
	N°	*%*	*N°*	*%*	*N°*	*%*	^{2}	*p*
Hitting with hand	1	10	7	28	20	41	4	.13
Object-hitting	0	0	3	12	20	41	11.19	<.01
Skin-picking	5	50	2	8	0	0	27.18	<.01
Self-scratching	2	20	0	0	7	14	4.55	.10
Grinding teeth-self-pinching	1	10	1	4	3	6	0.46	.79
Object-finger in cavities	2	20	3	12	17	35	4.63	.09
Self-biting	1	10	3	12	19	39	7.69	.02
Hair-pulling	3	30	1	4	3	6	7.07	.02
Nail pulling-eating	4	40	6	24	6	12	4.71	.09
Teeth grinding	0	0	8	32	4	8	9.57	<.01

6. Conclusions

The data reported indicate, in reference to PWS, that in our sample the SIB with the most frequent occurrence is skin-picking (50%) with similar results in other studies (Symons *et al.,* 1999) even if our percentages result lower. Wigren and Hansen (2003) have reported in their research, besides skin-picking, pulling hair as a SIB characteristic frequent in subjects with PWS. In our sample this behavior is present in 40% of cases. Other studies (Dykens *et al.,* 1999; Dimitropoulos *et al.*, 2001) have shown the presence of the behavioral characteristics referred above. Concerning body location of SIB, our data confirm the topographical areas most frequently involved as the hands, legs, arms and head.

In our sample with DS the SIB most frequently results in teeth grinding, eating nails, body-hitting with the hands. Our data, in this case, are not in agreement with that reported by other authors.

Fisher *et al.* (1998), Hagopian *et al.* (2005) and Määttä *et al.* (2006) have shown that in subjects with DS, when SIB is present, more frequent characteristics are eye poking, hitting and turning the ears with the hands. This result is not confirmed by our sample. Regarding bruxism and onicofagy, behaviors about which literature is discordant on whether to consider them self-injury behavior, our opinion is to consider them in a broad definition of

SIB, generally understood as behavior that causes or could cause physical harm.

Symons *et al.* (2003) and Canitano (2006) in their studies indicated "self-biting" and "hitting oneself" as SIB more frequent in AD. Our data are in only partial agreement with this, as far as considering this behavior present respectively in 39% and 41% of cases reported, while a similar percentage (41%) is reported for body-object hitting, and 35% for "finger/object in cavities".

Relative to specific body areas, as in the previously cited studies, also in our sample there is a prevalent involvement of the hands, head, face, and arms.

In literature it has been reported that an insurgence of SIB occurs frequently during early childhood (Oliver *et al.*, 1987).

Our analysis has shown that the range of age in which SIB emerges coincides in all three syndromes, and may be identified in the age range between 0 and 6 years (90% for PWS, 72% DS, 72% AD). The same coincidence between syndromes has been shown for the range from 7 to 12 years (10% for PWS, 12% DS, 10% AD).

To explain differences, we should consider that a wide-range definition of SIB reveals a series of behavioral symptoms that are not unanimously agreed-upon by researchers as signifying SIB. Some of them could be considered part of other psychopathological conditions, as for example the obsessive-compulsive spectrum.

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