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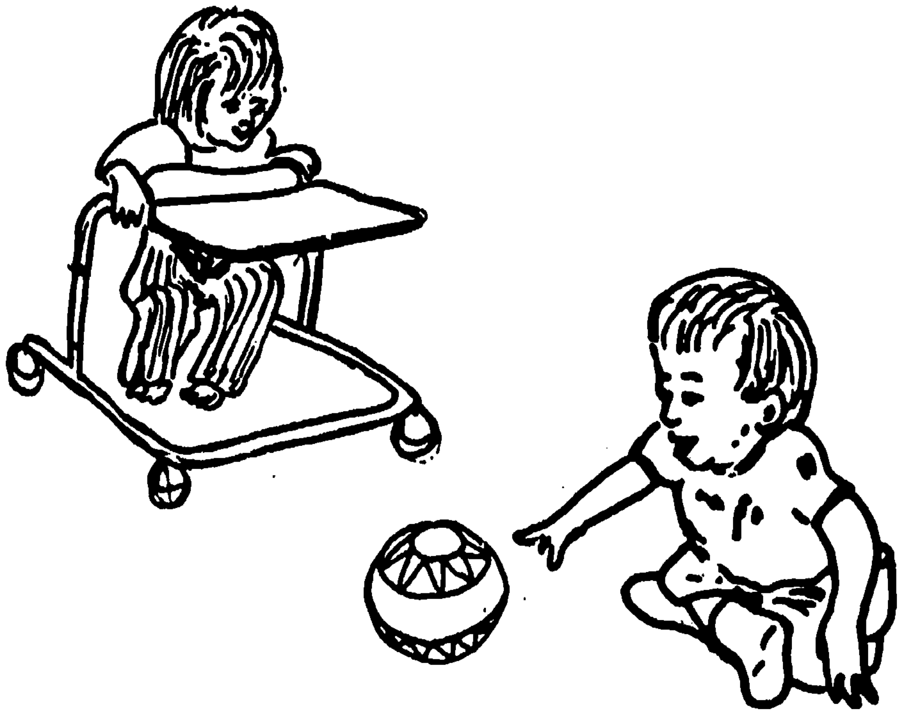
ABSTRACT

Information is presented for caregivers of Swedish children with osteogenesis imperfecta (brittle bones) and their families. Approximately five children with brittle bones are born in Sweden annually. Two main types of brittle bone disease have been identified: congenita and tarda. Typical symptoms include numerous and unexpected fractures, bluish colored eyeballs, brittle teeth, protruding rib-cage, small stature, and in some cases, hearing impairment. There are inadequacies in the support that the government provides for families with osteogenesis imperfecta children. Various kinds of support available include rental and home assistance, home adaptation allowance, child care allowance, respite service, transportation assistance, subsidies for drugs and medical care, and technical aids. Habilitation services include toy libraries, preschool and leisure time centers, and the school system. Case studies of two preschool children are presented, based on family interviews, and are illustrated with photographs. A list of four suggested readings and four related readings is included. Brief descriptions of The Society for Disabled Children and Adolescents (Stockholm, Sweden) and WRP (Women Researchers in Play and Disability) International conclude the document. (JDD)

JOHANNA AND TOMMY

TWO PRESCHOOLERS IN SWEDEN WITH BRITTLE BONES



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ABSTRACT

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JOHANNA AND TOMMY - TWO PRESCHOOLERS IN SWEDEN WITH BRITTLE BONES

29 pages

The aim of this study has been to work out an initial information for parents and caregivers and all those who come in contact with children with brittle bones and their families.

The study is based on interviews with two parents with preschoolers with brittle bones and photos of two of the children suffering from this disease.

Key words: Osteogenesis imperfecta, brittle bones, preschool children

Translation: Christina McCarthy, November, 1990

FOREWORD

This booklet you now have in your hand was created by WRP International in co-operation with the RBU group that works with OI (brittle bones). We hope it will be enjoyable and serve as initial information for parents and care-givers and all those who come in contact with these children and their families. We also recommend the project report "Pre-schoolers with brittle bones - psycho-social support for families with a pre-schooler who has brittle bones", to those who would like a more in depth knowledge of OI and the situation in Sweden. That report can be ordered from WRP.

A heartfelt thank you goes to Johanna and Tommy and their families for taking part in bringing about this booklet. We also want to thank The Swedish State Inheritance Fund which financed this project.

WRP International	RBU
Jane Brodin and Kristina Millde	Anita Engström

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INTRODUCTION

Each year approximately five children with brittle bones (latin name: osteogenesis imperfecta, OI) are born in Sweden. The knowledge about and the experiences from this disease are very limited due to the group's small size.

Many parents feel that it is important to receive information at an early stage about the disease and how it will affect their child's future development. The parents have the right to proper information which should be adapted to the individual family's abilty to receive and understand it. Several meetings with the parents are often required since they have difficult news to acquaint themselves with. It is important to allow the parents to arrive at and express their own questions and expectations and the support should be adjusted to these needs.

To date there is not much written about children with brittle bones neither in the Scandinavian countries nor in any other country of the world. Descriptions of "porcelain people" from early history tell us that this disease is very old indeed. Medical research has questioned earlier treatment methods and resulted in the development of new ones. The real solution to the puzzle has not yet been found. A cure does not exist and the effect of various treatments now in use is doubtful.

WHAT ARE BRITTLE BONES?

Within the work group for OI, RBU (The Society for Physically Disabled Children and Adolescents) has proceeded from two main types of brittle bone desease: congenita and tarda.

Congenita is the least common and the child with this type of OI is born with pronounced symptoms of the disease. In some cases the illness is so severe that the infant dies already during the fetal stage or shortly after birth. Many children are born with unhealed or improperly healed fractures.

Tarda appears later - sometime between the age of six months and two years. There is a milder and a more severe form of Tarda, the latter type resulting in many fractures and severe deformations of the limbs.

There are different types of OI and to merely divide the disease into two types is in reality not adequate. Within each main group a number of variations exist depending on the degree of severity of the disease and when it appeared. For this booklet, however, we have chosen not to become too technical but merely want to mention the existence of many more types of OI. One can, however, recognize a division into two main groups, Congenita and Tarda.

The following are some typical symptoms of brittle bones:

- numerous and unexpected fractures, i.e., as a result of a light bump or even a quick muscle movement
- blueish coloured eyeballs
- brittle teeth
- protruding rib-cage
- small stature
- in some cases,later on, hearing impairment

The various types of brittle bones have one thing in common: the incorrect structure of a protein known as collagen.
Normally, collagen serves as a reinforcement of the bonetissue. This function is reduced with the change in structure. OI is hereditary but occasionally it appears in a family with no previous record of the disease. The reason for this might be a latent disposition towards the disease or changes in the genes

by mutation.

The OI Society of Canada has made up a card which must accompany the child just like the Swedish patient cards. The card reads:

BRITTLE - HANDLE WITH CARE!
I have OI and my bones break very easily.
You must:
NEVER push, pull, bend, apply pressure on or try to straighten my bones
AVOID turning my head
AVOID raising my legs when changing my diaper
ALWAYS place your hands under my behind and slide in the diaper
ALWAYS handle me carefully, with careful movements, support my head, body and behind
ALLOW my parents to be present during X-ray and other examinations

They know how to handle me

This can be very useful when in contact with people who do not know how to handle a child with OI.

Technical Aids

There are great inadequacies in the support that the state provides for families with OI children. This is apparent from a study done by the Swedish Handicap Institute. Many families' needs for technical aids are badly looked after or not taken care of at all. The lack is apparent in several areas:

* the body providing technical aids moves exceedingly slowly
* poor information about available technical aids
* development and adjustment of the existing aids is needed

The only way to accomplish improvements is to keep on demanding a change. Many families who need their home adapted solve these problems themselves, unaware of the fact that the state has special offices responsible for these tasks.

Many OI children are rather lonely and have few friends. This is probably due to the fact that the parents are worried about the risks for fractures. Playing with other children, however, is an important part of the child's development whether he/she is healthy or not. If the parents allow their concerns to direct the child's life too much they risk hampering his/her development. The parents must, however, discover for themselves when and to what extent they can let go of their child. With maturity and growing awareness of the handicap the children and their parents arrive at their own way of managing.

Families with OI children are often subjected to suspicion of child abuse when they arrive at the hospital emergency department and the child has a fractured bone. It is, therefore, important that the diagnose can be made as early as possible in order that these kinds of experiences might be avoided.

In the next chapter you can read about the various kinds of support provided by the Swedish state and where to go for help.

SUPPORT FROM THE SWEDISH STATE FOR PHYSICALLY DISABLED CHILDREN

Below is a short overview of the various kinds of support available, how to go about requesting help and where to direct your inquiries in different matters.

FINANCIAL AND PRACTICAL HELP

Rental and home assistance

The approval of the rental and home assistance allowance depends on the family income and is paid out in very specified, given situations. The social welfare office looks after this.

Home adaptation allowance

If your home is of acceptable standard you can apply to your municipality for home adaptation, i.e., ramps, removal of thresholds etc.

Child care allowance

You can get a child care allowance for a child under the age of 16 who lives at home and requires more than the normal amount of care. The size of the allowance depends on various factors. This information can be received from the the health insurance office.

Respit service

Some municipalities have home- or child-care workers who can give the parents time off from taking care of disabled children. Besides this service there is also respit service according to the Social Welfare Law. Inquire at the social welfare office.

Transportation assistance

The social welfare office has the information about the rules in force for this service in your municipality.

The 15 - card and the gratuitous card

These cards are used for drugs and medical care. If the child requires frequent doctors appointments and continuous drug treatment these cards will cut the costs. Information from the health insurance office.

Technical aids

In Sweden everyone gets, as a rule, the technical aids he/she requires for free or on loan for as long as they are needed. Go to the municipal health nurse's office of BVC (Swedish child health care centre) for information.

HABILITATION

Each county has an office with the specific function of giving medical, educational and councelling support to families with a disabled child. This function is called habilitation and may vary depending on each individual child's disability. Direct your inquiries to BVC (The Swedish Child Care Centre) or a childrens' clinic; they have all the information.

Toy library

The toy library (lekotek) is nowadays often incorporated with the habilitation and can be of great value to disabled children and their development. Here you can get individual advice and practical training.

Preschool and leisure time centre

From the social welfare office you can get the information about the possibilities of getting in to one of these facilities with the required adjustments which must be made for your child, i.e., additional staff, the size of the group, modification of the building.

The compulsory school system

The municipal board of education is responsible for making it possible for each child to attend a school by providing certain services, i.e. a personal assistant, technical aids, modification of the building.

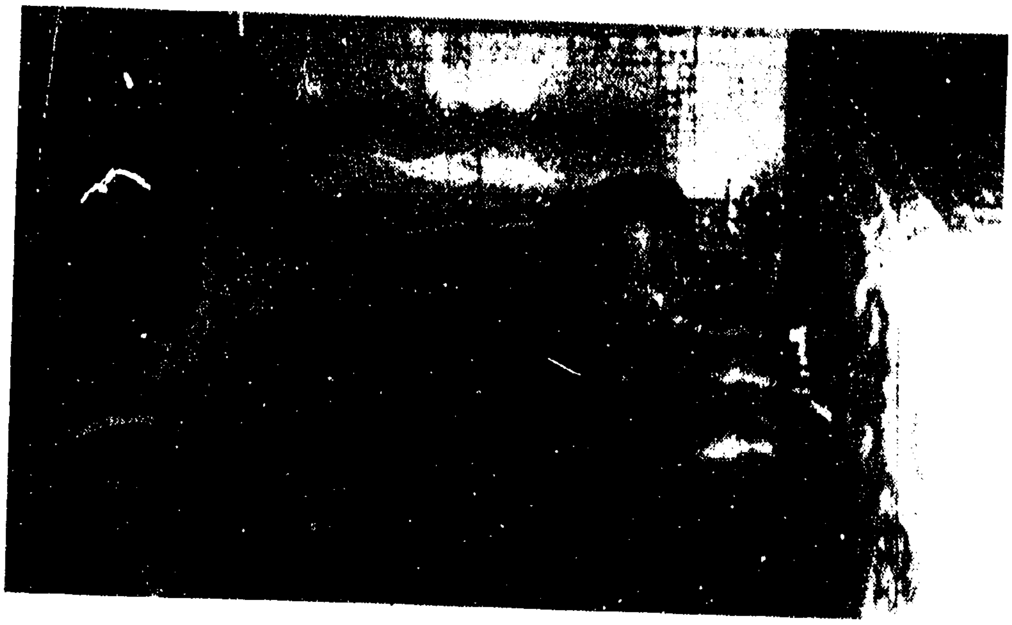
In general terms, the above is information about the disease and the assistance provided by the state to the families. In order to give you a more specific picture we now present Johanna and Tommy, two children with brittle bones. You will get acquainted with them and their families. The parents told us during the interviews how they felt initially after their child with OI was born and what kind of help they have received, both medical and practical.

Johanna's illness was diagnosed directly after birth. She was born with fractured arms, legs, collarbones and skull. The doctor realized immediately what the problem was and told them that he suspected OI. The following day an X-ray confirmed his suspicions. When the parents received the news they were mainly worried over the possibility of brain damage. The next day they met an orthopedic surgeon who gave them complete details. Since then they have almost always had the same doctor. This feels good and safe and the mother mentioned several times how happy she is that Johanna was born in Stockholm where all the facilities are near by.

[redacted]

From the age of four months during approximately a six month period Johanna received injections of Calcitonin three times per week. This treatment was discontinued since it caused nausea and vomiting. Johanna has routine check-ups, a hearing check and a visit to the dentist four times a year. These regular check-ups are important because dental and hearing problems often go hand in hand with this illness. Except for the above mentioned, they only visit a doctor when required.

This family has not had the experience, which is quite common, of being suspected of child abuse when consulting a doctor. They are, however, sometimes treated with suspicion by the hospital X-ray staff who do not listen to the parents when they explain how Johanna must be treated. "I wish they would trust me", the mother says. When Johanna has a fracture she must be taken to the emergency. Sometimes the mother phones ahead to find out who is on duty there. If it is a fractured arm they prefer to wait and see their regular doctor. It is important to see the right people and avoid lengthy explanations in an emergency situation.



The family has been in touch with a welfare officer from the beginning and this is working out well in practical matters. The mother is a physiotherapist and very knowledgable and also knows how to get in touch with the proper authorities etc. She has managed to arrange a care allowance from the health insurance, a further allowance from the social welfare office and also a municipal home assistance payment. They have not had any problems receiving the financial support which they are entitled to from their municipal offices. The mother has arranged physiotherapy and exercising in a pool. She prefers that someone besides a family member looks after the swimming according to a pre-arranged schedule. She only wants to be Johanna's mother, and not her physiotherapist as well.

The family receives thirty hours per month which are paid for by the municipality in respit service and baby sitting. At first they had a young girl who was to learn how to look after Johanna, but this did not work out. Johanna became upset. The mother's sister was a frequent visitor and finally they asked that she would become the paid baby-sitter. This was eventually arranged. In principal the municipality is willing to approve more hours when required. "You must always find the information yourself. The official offices hold back on certain services we are entitled to". At least that is how it sometimes appears to the people concerned. Much is lost in the flood of information and it appears that it was never given out. The family was offered the assistance of a psychologist already when Johanna was in the newborn ward but they declined. They have had much support from family and friends. The parents have worked themselves through the crises at home. We can see that the family has received both medical and social support. Their only complaint is the treatment they receive in the emergency ward. The ignorance of the staff there causes great concern and they also display an apparent lack of understanding. Parents, brothers and sisters and others around them have done their best to be understanding which has meant a lot to this family.

The mother has been participating in a mothers' group ever since Johanna was four months old. This has been mainly a positive experience. There were no problems as long as the children were little and stayed put, but when the others started to sit up and later learn to stand she became upset. At that time Johanna's handicap became more visible and that is also when the concerned calls started, both to the playmates and to herself, to be: "careful with Johanna, careful with Johanna".

Already from the beginning the family has had good doctors who could give them proper information. The mother asked already the first day on the orthopedic ward if there was an organization for children with OI and filled in the application form. She was then contacted by them. From then on, the best support came from other OI childrens' parents who already had the knowledge and experience. When Johanna was four months old the whole family went to camp where they made many valuable contacts through which the mother became fairly well informed about what OI entails. Before Johanna was born the mother had met a girl with OI whose development was going well and she therefore felt fairly confident that Johanna would manage as long as there was no brain damage. It was more difficult for the father who only received information through the mother and had no other experiences to refer to.

Johanna is very much afraid of other children who move around making loud noises. She is starting to approach other children in the play ground, but keeps a distance. She uses her walker a lot at home where she can move about freely through the apartment since the thresholds have been removed. Her mother has noticed how she is developing faster now and how her comprehension is increasing. Earlier, she was not able to take part in poking about in the bathroom and in cupboards and drawers like healthy children do. The rooms have different dimensions now that she can move about by herself.

Johanna visits the toy library (lekotek) where she can borrow things and through playing with these toys she learns about the world around her. Sometimes she uses these things differently from what was originally intended. Johanna is a sweet and happy little girl and is developing normally but at a slower pace.

INTERVIEW WITH TOMMY'S FAMILY

Tommy is six years old and lives with his family; mother, father and an older sister, high up on a hill in a house which they built themselves. The hilltop is not a convenient location for a disabled person.

Tommy's OI is of the mutation type, which means that it was not possible to detect earlier that the disease, which is hereditary, was in the family. The birth was by Caesarean section and the doctors realized immediately that something was wrong with the baby. His head was soft and one thigh bone was crooked from a healed fracture. Tommy's arms also had healed fractures. He was transferred to the newborn ward in a different hospital and placed in an incubator. The mother wanted to accompany him there but hospital routines prevented this. Thanks to a competent and resolute nurse she was able to go there the next morning. Everything was confusing and disorganized in connection with the birth and also during the days directly afterwards. The parents were to get all the information at the hospital from a team of doctors; the orthopedic surgeon, the pediatrician, the assistant physician and a few others. They all arrived late for the meeting and rather cooly advised the parents that Tommy had brittle bones.

Tommy stayed at the hospital for approximately three months after delivery. The mother had his sister, 18 months old, at home and was worried about how she would be able to take care of Tommy. The staff at the hospital tried to pursuade her to take him home but she did not dare to. At Christmas time they took Tommy home on a trial basis which turned out well and he was able to stay home. From the age of three during a period of one year Tommy received injections of Calcitonin. There was no noticable improvement and the treatment was discontinued. Tommy has not had many severe fractures. Often the mother can take care of him herself when he gets hurt. When fracturing his arms or legs, however, they must go directly to the emergency. At one time when his thighbone required to have a nail inserted he ended up with another fracture during the surgery. More operations! This in spite of the fact that the surgeon was the one with the most experience with OI. Tommy is exceedingly brittle.

In the beginning the parents had certain problems with rough staff in X-ray departments and an orthopedic surgeon who pulled and poked him. To him, Tommy was an orthopedic case but of no interest as person. A doctor who believes in natural medicin in Malmö suggested three different natural medicines which have had a very good effect on Tommy's general health and resistance to infections. His joints now also ache less and the mother believes this is due to these natural medications.

Tommy went to a habilitation team when he was a year and a half old. The staff there were very knowledgable and also compassionate. Tommy started going to a daycare centre when he was three. This has been a great benefit for his social development. "It's the best thing we could have done for him", the mother tells us. Tommy has a resource person there. The staff at the day care centre got their information from the mother and to a certain extent from the habilitation team. Most of their friends and aquaintances keep away or make light of the family's problems. Even their relatives seem to have trouble understanding what it is like to live with OI. The paternal grandparents have been babysitting a fair amount and lately even the other grandmother. For a while they had a "support family". Their relatives did not understand this arrangement. "One ought to look after oneself", they say. They actually cannot comprehend that this family needs more support than other families with healthy little children. That the family has received financial assistance and had a support family has actually caused a certain amount of envy among the relatives and neighbours.

It is easiest to socialize with people who are in the same situation. The other OI children seem like brothers and sisters to Tommy. The mother has asked the staff at the hospital to call her when an OI child is born. She very much wants to help other parents. From her own experience she knows how lonely you are when you first get the bad news. To have someone to talk to who really understands is very important and helps you get through the first crisis. By spreading information in various ways she hopes that more people will accept Tommy and the other children with OI. Their self-esteem must be protected. The family is planning to build a new home, this time not on a hill but on flat terrain, where Tommy can drive in and out of the house in his powered wheelchair and move about as he wishes. It is important that he becomes independent before he starts school.

The mother felt that the hospital staff did not show much respect or compassion after Tommy's birth. One of the people who was to supply them with information about Tommy's illness asked: "Why do you sit here crying all the time, can I do anything?" Such a remark shows how insensitive a doctor can be but other staff members have often displayed a better ability to understand and be supportive. The staff at the habilitation unit is good. They are wonderful to Tommy and his mother. Once a supervisor from the team visited them at home and spoke to them in a straightforward way about crisis situations and how to deal with them. That was the first time anyone spoke to them about emotions and crises which become part of life when you have a disabled child. Doctors at the hospital often talk above Tommy's head although he very well understands what is going on and can speak for himself.

Tommy will manage well socially because he is an intelligent, cheerful and "peppy" boy. With aids like wheelchairs and various modifications he will be able to lead a relatively independent and pleasant life. He will, however, require a certain amount of assistance because of his short stature.

Tommy can move about by himself on the floor and participate in games as long as they are not too rough. He moves around faster between places and activities in his wheelchair. Tommy's limitations stem from the fact that he is very small. One personal assistant at the daycare centre has provided all the help he requires. Later on in school and at the leisure centre he will also have extra staff people.

Tommy's mother is the area contact person for children with OI and now has a rather clear picture of what OI entails but the process of reaching this point was a difficult one. Most of her knowledge comes from her own studies.

CONCLUSION

This booklet is intended as initial information to parents, staff, relatives and all others who are in contact with children with brittle bones in Sweden.

The report: "Preschoolers with brittle bones - psycho-social support for parents with preschoolers with brittle bones" give a more detailed description and history of more children and their families.

We hope that together these publications will help those who need more knowledge about a rare disease.

SUGGESTED READING

Brodin,J. (1989) Barn med benskörhet och deras livssituation, Handikappinstitutet, CM Gruppen, Bromma. (Children with brittle bones and their situation in life, The Swedish Handicap Institute, CM Group, Sweden).

Brodin, J. & Lindberg, M. (1988) Avlösarservice - en rättighet eller ett privilegium? (Respit service - a right or a privilege?) WRP International, Samhall Kalmarsund Repro, Sweden.

Brodin, J. & Millde, K. (1989) Förskolebarn med benskörhet. Psykosocialt stöd till familjer med förskolebarn med benskörhet, (Pre-schoolers with brittle bones. Psycho-social support for parents with pre-schoolers with brittle bones), WRP International, Samhall Kalmarsund Repro.

Ekstrand, G. (1986) Barn med OI (benskörhet), Informationsskrift från Riksförbundet RBU, (Children with OI (brittle bones), Information booklet from RBU), Stockholm, Sweden.

RELATED READING

Barn och ungdomar med handikapp, Allmänna Råd från socialstyrelsen, (Disabled children and adolescents, General advice from the Board of the Social Welfare Office), 1985:9, Stockholm, Sweden.

Hvordan kan vi hjelpe? Norsk Forening for Osteogenesis Imperfecta, 1982 (How can we help? Norwegian Society of OI).

Rörelse nr 4/85, Tidskrift från RBU, (Movement no.4/85, Publication from RBU), Stockholm, Sweden.

Samhällets stöd till barn med handikapp, Allmänna Barnhuset och Statens Handikappråd (State support for disabled children, General Children's House and the Federal Handicap Advisory Board).

(RBU) THE SOCIETY FOR DISABLED CHILDREN AND ADOLESCENTS An organization for children and youth with physical disabilites

The Society for Disabled Children and Adolescents is a non-profit organisation with 29 local associatinos around the country. The members are disabled youth and their parents and also others who support disabled peoples' cause in society.

The organization includes the following groups: brittle bones, CP (Cerebral Palsy), extreme shortness, MBD (minimal brain dysfunction), muscle disorders, Prader Willi and Spina Bifida. Each group has its own central workgroup and liaison officer in the clubs. Specialized workgroups guard the interests of on the one hand the people with multiple disabilities and on the other hand the situation of the disabled youth in the workforce and also look after education, leisure time and independent living arrangements.

The organization's secretary, phone:08-246265, handles the communication with workgroups liaisons.

If you want to write them, the address is:

Riksförbundet för Rörelsehindrade Barn och Ungdomar (RBU)
David Bagares gata 3
S 111 38 Stockholm
Sweden

WRP INTERNATIONAL (Women Researchers in Play and Disability)

WRP International was founded in Toronto, Canada, in the Spring of 1987. WRP is an organization which works with matters involving play and children with disabilities. WRP is a cross-discipline project-and researchgroup with expertise both in research and practical activities with disabled children. The goals of WRP are:

* to promote international co-operation among women working with disabled children

* to promote and support disabled childrens' possibilities to play in various ways and environments

* to work towards raising the status of play

* to stress the importance of play for disabled children through projects and publications

* to stress the importance of the child's own play activity

* to support parents of disabled children by stressing the importance of play and interaction

* through research and development stimulate progress in the field of play

WRP has from the beginning carried on a number of projects resulting in publications mainly for parents, care givers and child care workers. These publications may be ordered from WRP International. Address: Turenäs Gård, 38030 Rockneby, Sweden.

INTERNATIONAL
WRP

WRP International (Women Researchers in Play and Disability) is an organization researching the filed of play and disability. WRP International was founded in Toronto in May 1987.

Publications from WRP International:

1. Kom och Låt Oss Leka, 1987, rev 1989
2. Avlösarservice - en rättighet eller ett privilegium?, 1988
3. Invandrarbarn med handikapp - en kartläggning, 1988
4. Lekotek i Sverige, 1988
5. Internationell forskning kring samspel hos barn som använder alternativ kommunikation, 1989
6. Barn med handikapp i förskolan, 1989
7. Adoptivbarn med handikapp - sju föräldraintervjue, 1989
8. Lek med Oss, 1989
9. Sensoriska handikapp och flerhandikapp, 1989
10. Förskolebarn med benskörhet - Psykosocialt stöd till familjer med förskolebarn med benskörhet, 1989
11. Johanna och Tommy - Två förskolebarn med benskörhet, 1989
12. Några invandrarfamiljers syn på lek och handikapp, 1990
13. Synen på lek och handikapp i olika kulturer, 1990
14. Play is for all - Toy libraries in an international perspective, 1990
15. Johanna and Tommy - Two preschoolers in Sweden with brittle bones, 1990

The publications can be ordered from WRP International, Turenäs Gård,
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