



**Follow-up programme for Cerebral Palsy**

**Annual report 2007**

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## Foreword

This is the second annual report for CPUP (A Follow-Up programme for children with cerebral palsy) as a National Quality Register.

CPUP started in Skåne and Blekinge in 1994 and the programme became a National Quality Register in 2005. During 2006 four new county councils joined the programme and from 2007 all of the county councils in Sweden are participating in the programme. Norway started to register in Helse Sør and Öst in 2006, which corresponds to half the population of Norway and our Norwegian colleagues are participating in the revision and the development of the programme. Denmark has also announced an interest to participate in the programme.

Cerebral palsy is a condition with a lower incidence than in many other conditions that are registered via the National Quality Registers. People with cerebral palsy are in need of regular follow-up and often needs life long treatment. Instead of monitoring the result of one given treatment to a vast number of individuals, CPUP monitors the results of continual treatment input to a lesser number of individuals from childhood up to adulthood.

CPUP also differs from most other registers by the fact that it is also used as a clinical instrument to follow a single individual, encompasses a structured chart, and also serves as a general follow-up, a quality register, for all children and young people with cerebral palsy. The information provided also gives us a unique opportunity to evaluate different aspects of cerebral palsy in a scientific way.

This second annual report is a presentation of all the collected results from the participating regions. Comparisons of treatment results between the different regions are not available as yet, as most regions have only started to register recently. Such analyses can probably be made within the next few years.

Transfer of reporting from paper charts and Filemaker to internet based reporting via the 3C system was a dominating task during the year. This has been done simultaneously to the revision of the contents of the occupational therapist and physiotherapist assessment forms, and also to the introduction of the neuropaediatric assessment forms. These changes have meant a lot of work for the people involved. The great alterations that have now been made in the programme cannot occur without some hindrances and difficulties occurring in the modification process and therefore we would like to take this opportunity to thank all of you who has helped us to go through with these changes, and at the same time has shown such understanding and patience. We have a lot of work ahead of us, both with developing the 3C system and the continued evaluation and development of the assessment forms. Hopefully, the greater part of this has already been done, so that we in the future can work more with smaller adjustments and modifications.

CPUP is built on the co-operation of all of us who work with children with cerebral palsy and is successively developed by tying together experiences from the professions involved in the care of the patients from different parts of the country. Once again, we would like to thank all of you who have contributed to CPUP and it is our hope that this report will provide you with feedback, thoughts and inspiration for continued good work and co-operation.

*Marianne Arner, Caroline Gudmundsson, Gunnar Hägglund, Henrik Lauge-Pedersen, Penny Lindegren, Eva Nordmark, Kerstin Sommerstein, Philippe Wagner, Lena Westbom.*

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## What is CPUP?

A follow-up programme, for children with cerebral palsy, CPUP, started in Skåne and Blekinge in 1994 as a joint project between the orthopaedic departments and child rehabilitation units. The reason for this co-operation was that we saw that a number of children with CP had developed hip dislocation and severe contractures. We wanted to prevent these complications by creating a new system in order to monitor the children in a structured way during their childhood and adolescent years. New treatment methods to reduce spasticity, such as botulinum toxin injections and baclofen pump treatment, increasingly used since the early 1990's, have shown to be more effective if used early.

The purpose of CPUP is:

- To prevent hip dislocation and severe contractures by continual and long-term examination of joint motion and if necessary, early preventive treatment input. One can then optimize function and heighten the quality of life for children and young people with cerebral palsy.
- To increase the knowledge of CP and the effects of different treatments.
- To improve the communication and the co-operation between the different professionals involved in the care of children and young people with CP.

The basis of CPUP is to identify children with CP and offer participation in the programme. The children are offered to join the programme at the initiative of their local habilitation teams, as soon as symptoms of CP are observed, i.e. even before a definite diagnosis of CP has been made, as hip dislocation sometimes occurs as early as in toddlerhood. With a recurrent systematic monitoring of habilitation/hospital patient registers and charts, additional children with CP or possible CP can be identified. After the age of four, the CP diagnosis and subtype classification can be determined. The children, who do not fulfil the criteria for CP will then be discharged from the programme.

Examination results are registered continually and feed back is given on a regular basis to the treating team so that adequate treatment can be given as soon as possible when deterioration is noted.

At the start of the programme, CPUP only involved examination of the lower extremities and the gross motor function ability. Since 2002, it also includes the children's hand and upper extremity function. In 2006, an assessment form was developed which provides information on the child's CP subtype and other impairments/disorders, which could have an impact on their motor development, for example visual ability and epilepsy, if any.

In 2005, we were able to present with a 10-year follow up, that hip dislocation is preventable and also that there was a decrease in the number of children that developed severe contractures in a total population. In 2005, CPUP received funding as a National Quality Register and in 2006, 15 out of 20 county councils corresponding to 86% of the population, have registered data in the programme. This year (2007) the five remaining regions have joined and have started to register patients. In Norway, registration takes place in two regions corresponding to half the population.

For more information: [www.cpup.se](http://www.cpup.se)

## **What is a National Quality Register?**

Today, there are 55 National Quality Registers in Sweden, which are supported by the Swedish Association of Local Authorities and Regions and The National Board of Health and Welfare. They have all been initiated by representatives from the medical professions and have been developed in order to enhance the quality of the clinical work. All registers contain patient information on diagnosis, treatment and results. This means that a follow up of what has been achieved in the health care system and an analysis of the differences between different hospitals and county councils can be made. The register fulfils the Health Board's confidentiality regulations. The Quality Registers are not intended for supervision.

The Swedish Association of Local Authorities and Regions and the National Board of Health and Welfare are co-operating on a central level and also contribute financially to the creating and development of the National Quality Registers. The National Quality Registers are therefore a collaboration project between the medical profession, the Swedish Association of Local Authorities and Regions and the National Board of Health and Welfare.

More information can be found on [www.kvalitetsregister.se](http://www.kvalitetsregister.se)

## **What is NKO?**

Today, there are three National Competence Centres in Sweden, of which NKO (The Swedish National Competence Centre for Musculo-Skeletal disorders) is one. The main assignment for the competence centres is to gather knowledge and create synergy effects by helping several registers with collection and analysis of data and statistical evaluations. CPUP is working closely with NKO in terms of the development of our webpage, development of our reporting function (3C), data adaptation and also statistical analysis for this annual report.

More information can be found on [www.nko.se](http://www.nko.se)

## Participating county councils and regions

As of January 2007, all of the county councils and regions in Sweden have joined CPUP. The regions which participated during 2005 were the basis of our first annual report in 2006. The table below shows the participating regions during 2006. The percentage number shows the proportion of the population aged 0-16, represented by each region. Initially, several regions chose only to participate in CPUP with regards to physiotherapist reports on the lower extremities and the hip and spine programme. In three of the regions, which last year only reported in the physiotherapist assessment form, the occupational therapists are now also involved in the follow-up programme. The regions who are participating in the entire programme have increased from five to ten. These are marked with an \*.

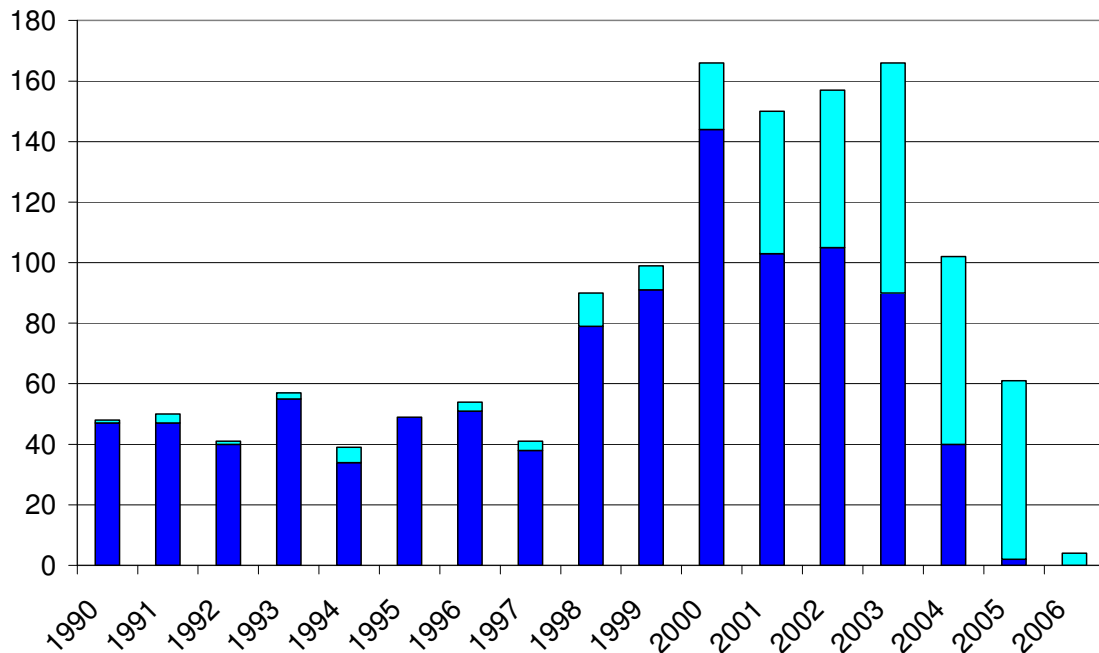
Participating regions 2006 (%)		New added regions 2007 (%)	
Skåne-Blekinge*	14.4	Gotland	0.6
Halland*	3.4	Norrbottn	2.6
Västerbotten	2.7	Jönköping	3.8
Västra Götaland*	16.9	Östergötland	4.5
Västernorrland	2.5	Kalmar	2.4
Stockholms läns landsting*	22.2		
Sörmland*	2.9		
Kronoberg*	2.0		
Dalarna*	2.9		
Örebro	3.0		
Västmanland*	2.7		
Gävleborg	2.8		
Värmland*	2.8		
Uppsala	3.6		
Jämtland*	1.3		

## Participating children/adolescents

As of the 31<sup>st</sup> December 2006, 1374 children were registered in the programme and they act as the basis for the material in this report (see figure). Of these children, 887 have so far also been reported in terms of arm and hand function.

Since the turn of last year, a total number of 359 children have been added to CPUP and this is an increase by 35%. It is very positive that so many of the new additions were very young, half of the children were under the age of three and one third was under the age of two. One of the great advantages of such an early participation in CPUP, is that it is one of the prerequisites for preventing hip dislocation.

The proportion of children who have been reported in terms of arm and hand function have increased with 235 (37%).



*Proportion of children in CPUP (N=1374) in relation to year of birth. Pale blue colour represents children, joining the programme in 2006. Children born in 1990-94 are exclusively from Skåne and Blekinge, from 1995 also from Halland, from 1998 also V.Götaland, Västernorrland och Sörmland, from 1999 also from Västerbotten, from 2000 also from Stockholm, Kronoberg, Örebro, Västmanland, Dalarna, Gävleborg and Jämtland, from 2001 also Uppsala and Värmland.*

Early intervention is a prerequisite for the programme. The children must be followed in CPUP, even before it is clear as to whether they have CP or not, as it sometimes can take quite a while before a proper diagnosis of CP can be made. CP is defined as a permanent motor disability due to a non-deteriorating brain dysfunction/injury before the age of two. There are a few children, who at an early stage displays signs of CP but where the neurological symptoms regress before the age of four. A few other children with functional impairment of a CP-type later appear to have a progressive brain disorder. These children should be followed in CPUP until it is clear whether they fulfil the criteria for CP or not. This is usually possible to determine at the age of four. The children, who do not fulfil the criteria for CP will then be discharged from the programme and monitored differently. Symptoms in children with a mild form of CP are sometimes not detected in the first years of life, but most children with CP have been diagnosed around the age of four.

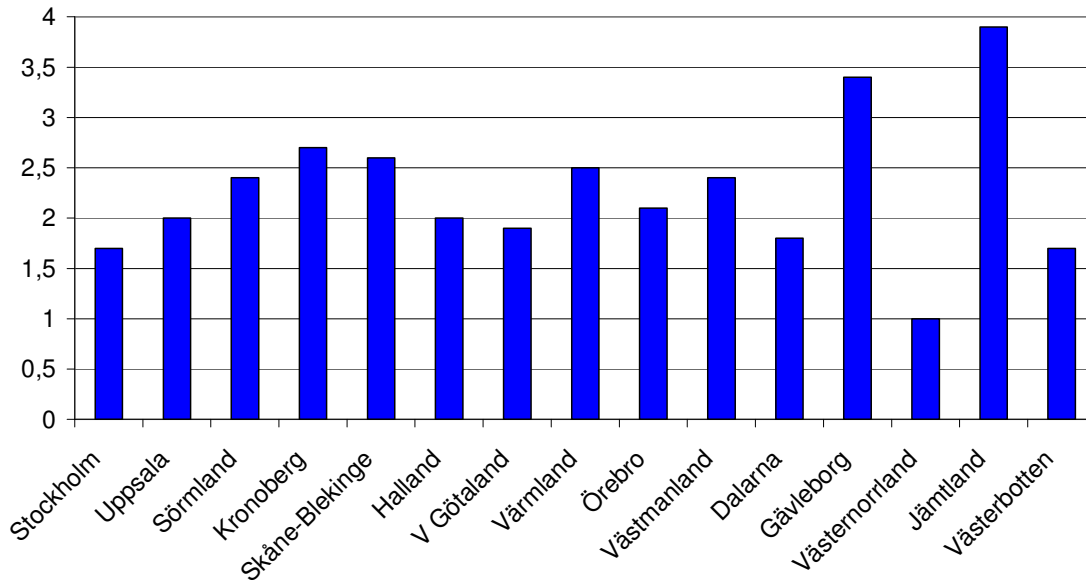
At the start-up of CPUP, most regions have chosen to include the five youngest age groups. Therefore, we have analysed the prevalence of CP at the end of 2006 for children born in 2000-2004, i.e. between the ages of 4-6.

At the turn of the year 2006/2007, 504 children in Sweden between the ages of 4-6 were being monitored in the programme. Spread out on all the participating regions this involves 2.0 /1000 children in this age group. This can be compared to the CP prevalence for the ages of 4-7 years in studies from V.Götaland (Western Sweden) 1.9/1000 (children born 1995-1998,



including only those born in Sweden) and in Skåne and Blekinge (Southern Sweden) 2.7/1000 (children born 1994-97, also including those born abroad) This supports the fact that most children with CP in this age group are being monitored in the CPUP programme.

**Regional prevalence of CP reported in CPUP**



*The proportion of children reported from respective region, born in 2000-2002 per 1000 children in the same age group in respective regions ( for Uppsala and Värmland the calculation is based on children born between 2001-2002).*

The prevalence of children, aged 4-6, who are being monitored in CPUP from each region vary according to the figure between 0.1 and 3.4/1000. The children in each age group are so few, even in the greater regions, that chance can explain the variances in prevalence number. The proportion of children with identified CP in respective region make up a quality measurement at the child health care clinics/habilitation centres. This and any possible differences in the real prevalence number between the various regions can only be discussed when more age groups successively are included in the follow-up programme.

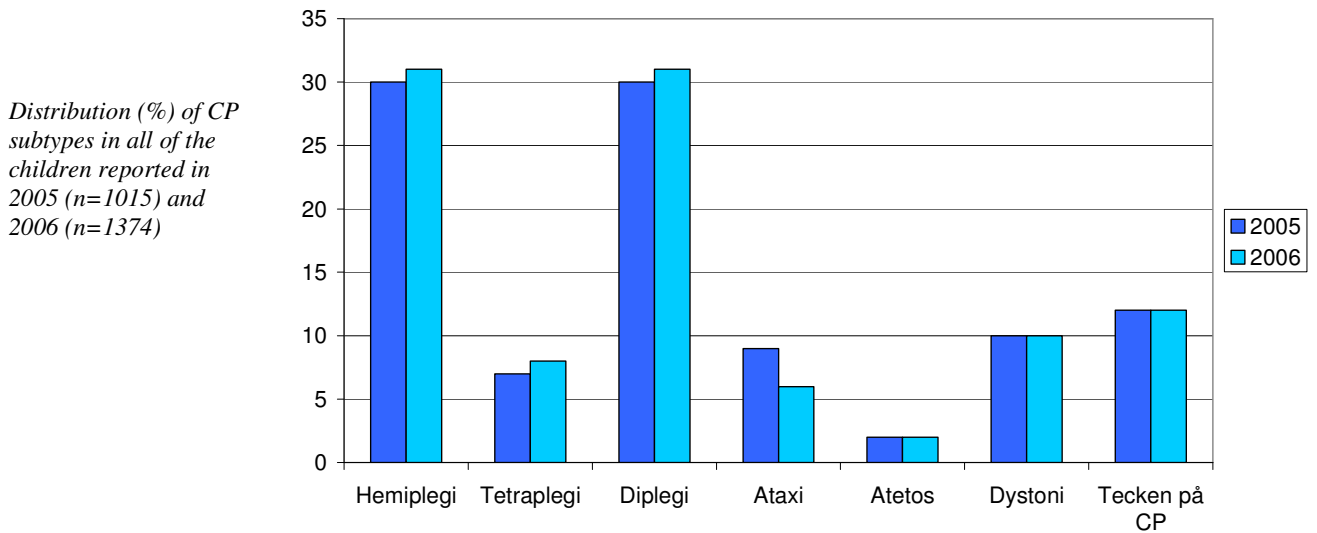
For the 1374 children participating in CPUP, we have received data from 6800 physiotherapist assessment forms regarding the spine and lower extremities, 2600 occupational therapist assessment forms have been received regarding the upper extremities and 2700 radiographic measurements of the hips have also been registered. Children under 6 years of age are examined at six month intervals and older children are examined annually by their physiotherapist and occupational therapist. The following report is based on information from the child's latest report/examination.

## CP subtypes

The distribution between different CP subtypes for the total material and the distribution between the various regions is shown in the figures below. The distribution nationally coincides rather well in comparison to previous epidemiological studies in Skåne, Blekinge (Southern Sweden) and V.Götaland (Western Sweden)

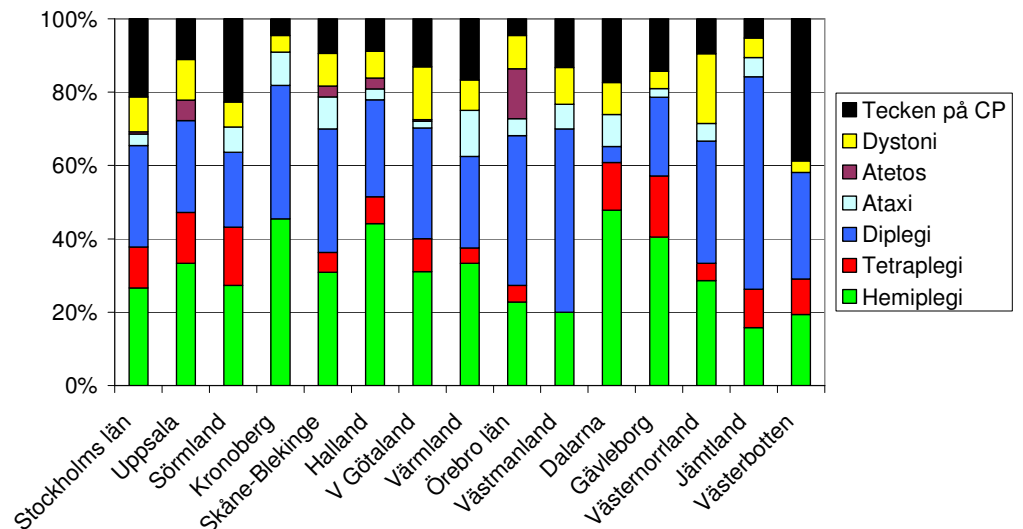
However, the distribution of CP subtypes varies substantially between the different regions. Some regions still have very few participants in the programme and the explanation to this may well be chance. It can of course also be a true difference in degree of severity between the various regions, or a difference in the evaluation of CP subtypes.

National distribution of CP subtypes (%)



Regional distribution of CP subtypes

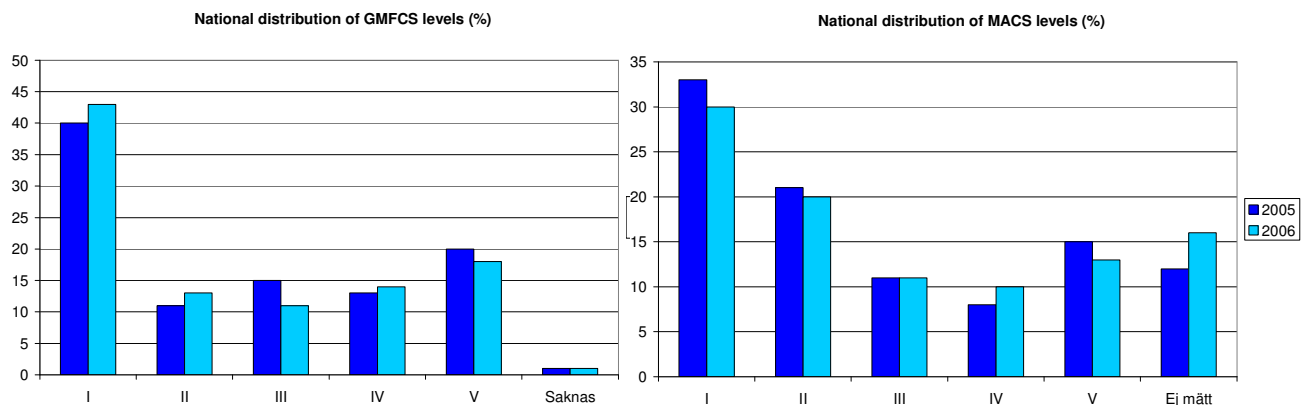
*Distribution (%) of CP subtypes for the total number of reported children (N=1374) distributed according to region.*



## Gross motor function and manual ability

Evaluation of gross motor function according to the Gross Motor Function Classification System (GMFCS) and manual ability according to the Manual Ability Classification System (MACS) is presented in the figures below. The national distribution of GMFCS levels indicates a somewhat higher frequency of children at level IV and V than previously presented distribution in Skåne and Blekinge. This may be explained by the fact that children, with a greater functional impairment are identified and diagnosed at an earlier stage. As several regions so far, only assess children in the age group 0-5 years, there will be an over representation of children at levels IV-V. This may also explain the trend of more children with less severe functional impairment.

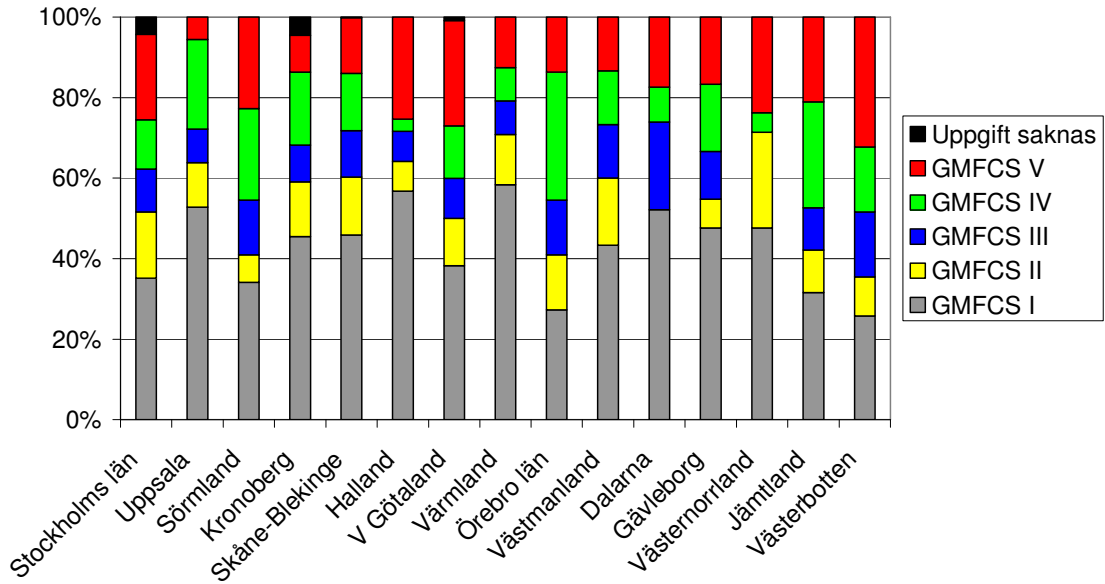
In 2006, MACS levels had been registered for 97 % of the children aged >4 years of age. The distribution between the levels was generally unchanged compared to 2005. About 70% of the children were noted as having “significant” problems with hand function (>MACS I), but half of them were still reported as being independent in their age-relevant manual activities (MACS I-II). Nearly every fourth child had a significant manual impairment with a total or almost total need of assistance in their daily activities (MACS IV+V).



*Distribution (%) of the number of reported children at GMFCS levels (N=1015 in 2005, 1374 in 2006) and MACS levels (N=563 in 2005, 775 in 2006). MACS evaluation is only carried out on children aged 4 or older.*

There is a large spread in GMFCS levels between the various regions. (see figure below) As for the analysis of CP subtypes, we must consider that several regions still only have a few participants in the programme, so the explanation for the above can be chance. The reason for the observed differences can both be variations in the number of children with mild CP, who have been identified and included in the programme as well as differences in determining the degree of severity of the functional impairment in the various regions.

**Regional distribution of GMFCS levels**



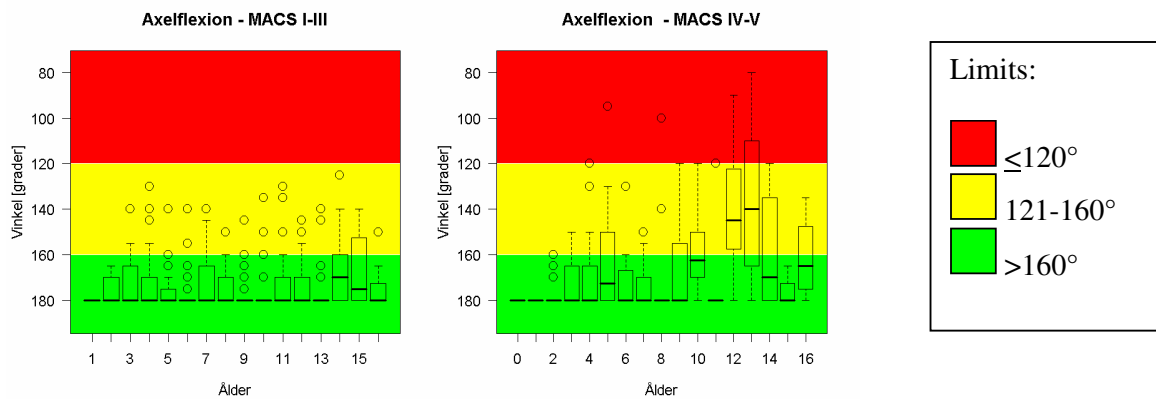
*Distribution (%) of the GMFCS levels in the total number of reported children (N=1374) per reporting region. Uppgift saknas=missing data*

## Range of motion

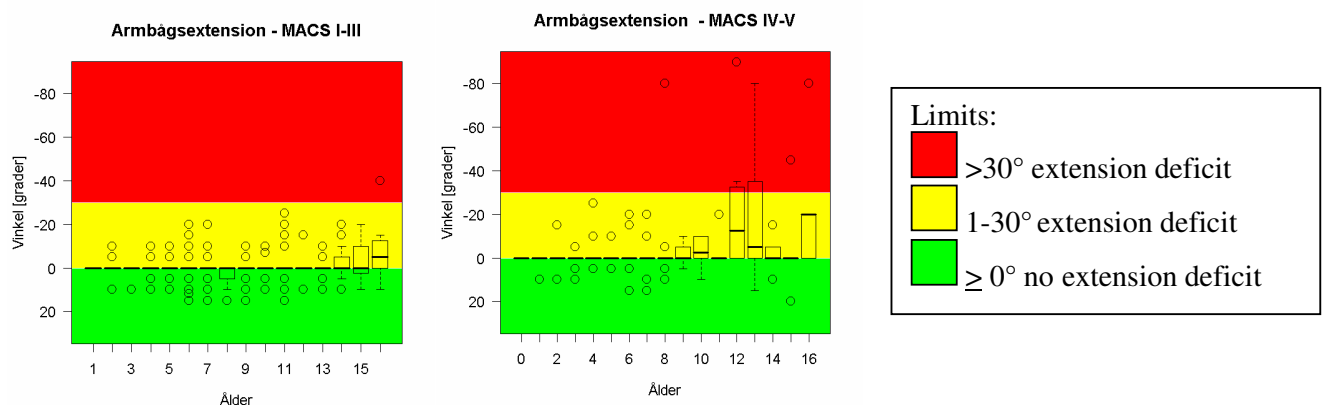
Shown below are the results of a selection of passive joint motion measurements divided into GMFCS/MACS I-III and GMFCS/MACS IV-V and age groups. The value from the most affected side of each child is reported. The passive range of motion measurements are illustrated with so called boxplots with median value and quartiles. Extreme values, so called outliers are marked as a circle.

In CPUP, we use limit values, so called warning lights of the traffic type model for each joint that is measured. Green indicates what we regard as a normal or almost normal value. Yellow value should, if measured correctly, bring about revision of the child's present treatment. A red value is clearly pathological. For the lower extremities, the limit values are different depending on the GMFCS level. The limit values in CPUP work on the supposition that the children should have adequate range of motion in the hip-, knee- and ankle joint to support proper standing (GMFCS level IV-V) or walking (GMFCS level I-III). As CPUP aspires to prevent contractures and deformities, the limit values are relatively narrow. The idea is that the occupational therapist and physiotherapist in charge, at the first sign of deterioration should be able to react, communicate and confer with the other clinical professionals involved. See also [www.cpun.se](http://www.cpun.se)

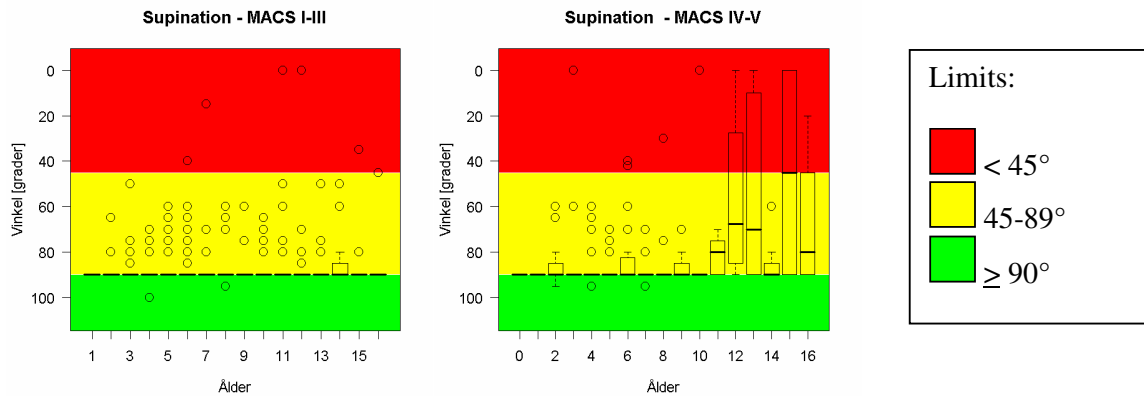
### Shoulder flexion/elevation



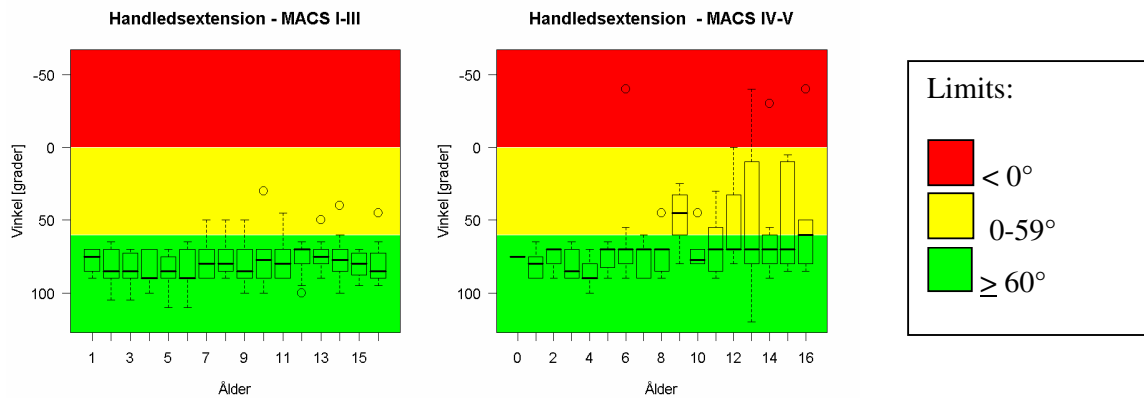
### Elbow extension



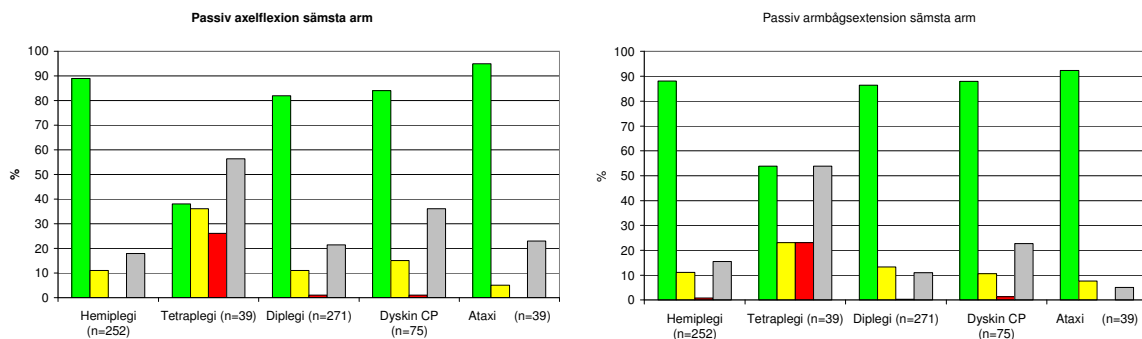
### Supination

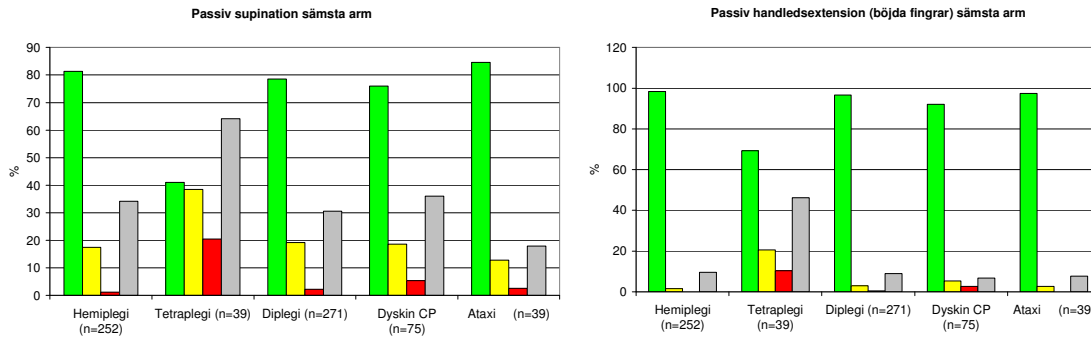


### Wrist extension



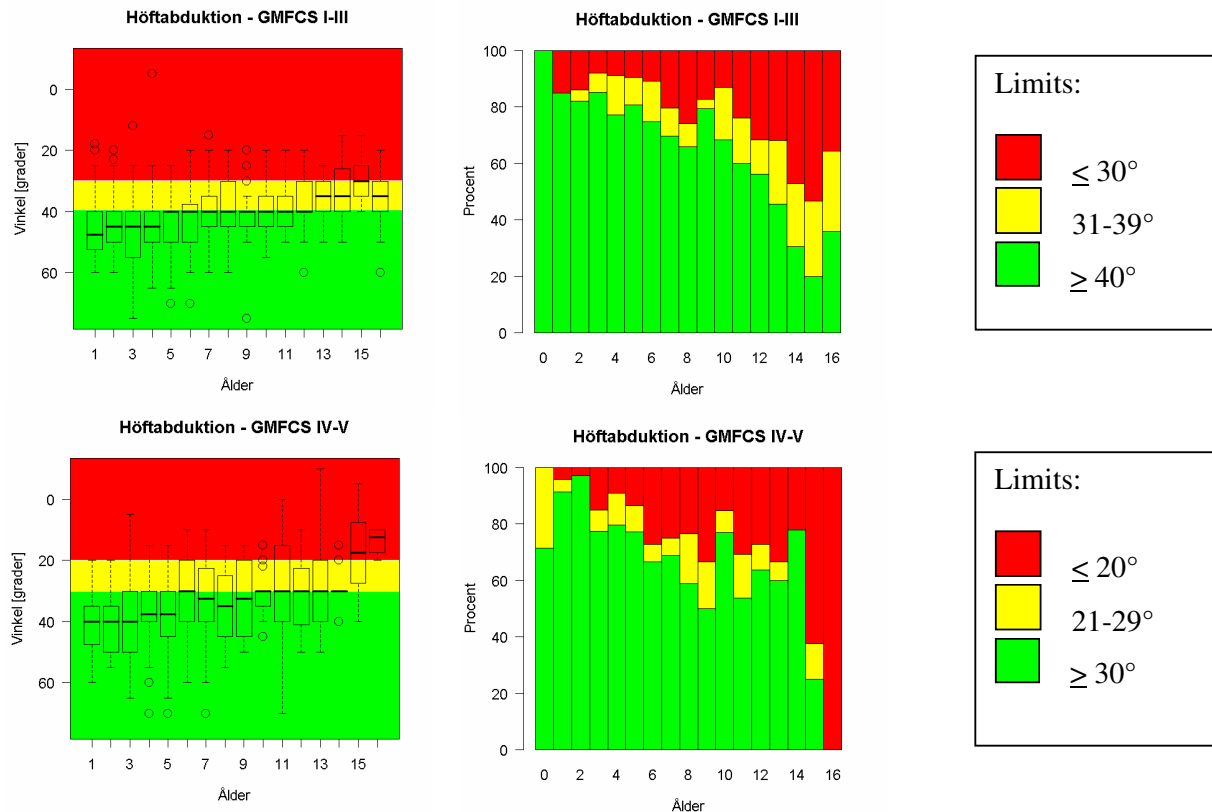
*Commentary upper extremities:* Data for all the registered measurements (on the 775 children who were classified at MACS-levels) is shown in the boxplots. We have chosen to divide the data into MACS I-III, i.e.. children who can use their hands fully or partially in their manual activities and MACS IV-V, which represents children with severe functional impairment in the upper extremities. The latter group consists of only 30%, please see figure on page 9. We can not, as yet, analyse development over time as both the higher and the lower age groups still are very young ( see figure page 17). MACS levels can not be classified accurately and definitely in children <4 years, but information still has been registered for some children.



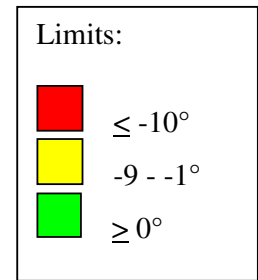
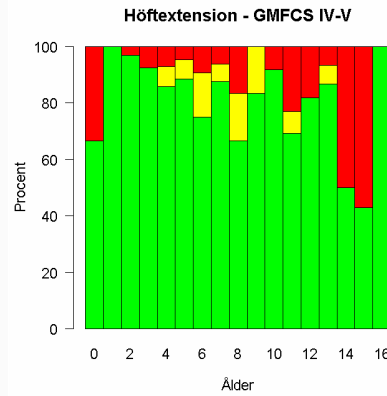
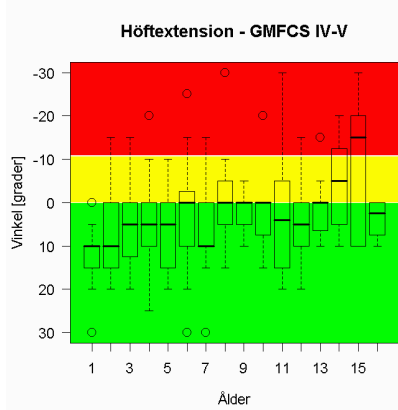
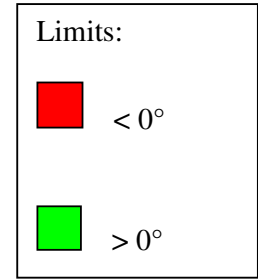
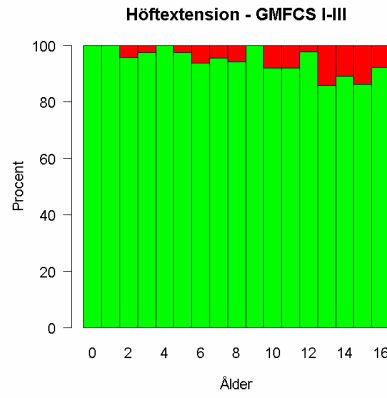
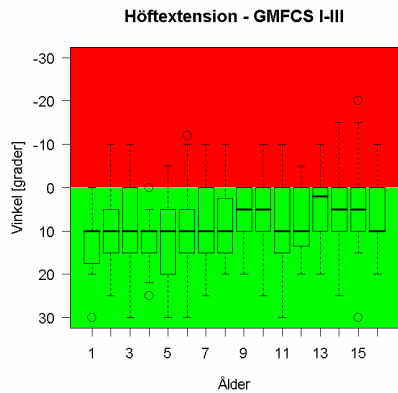


Above is shown in percent (%) the green, yellow and red values within the different CP subtypes and also the percentage, where tightness at passive motion has been noted (grey bars). Restricted passive joint motion (yellow and red values) was noted in shoulder flexion, elbow extension and supination in 40-60 % of the children with tetraplegic CP. Pronounced contractures (red values) were uncommon in the other CP subtypes. Yellow values were equally common in all the different CP subtypes (10-20%). These findings strengthen our ambition within CPUP to monitor *all* children with CP, irrespective of subtype. Just like last year, remarkably few yellow and red values were registered for wrist extension. It is difficult to measure joint motion in children with spasticity and some error values are possible. However, further education and training will gradually decrease any source of error value. There might also be a need to re-evaluate limit values for the different joint motions, in order to detect any possible movement restraints and tendencies towards contracture as early as possible. The focus on joint mobility, which CPUP has introduced, has already shown to be positive and it is our hope that resources are available so that adequate treatment input can be activated when a muscle contracture is detected. Not until then, is it possible for us to fulfil the goals for the programme.

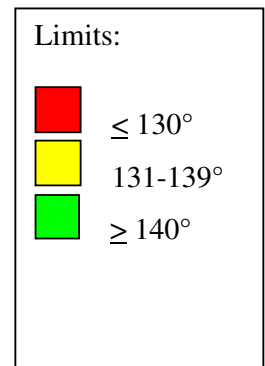
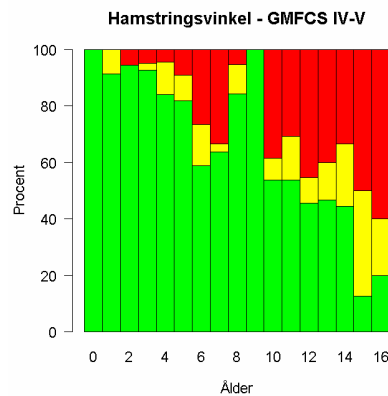
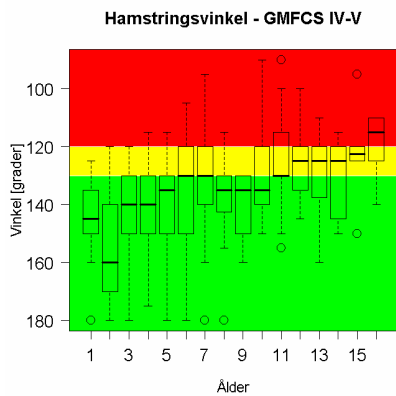
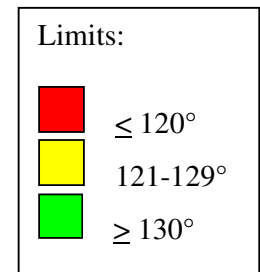
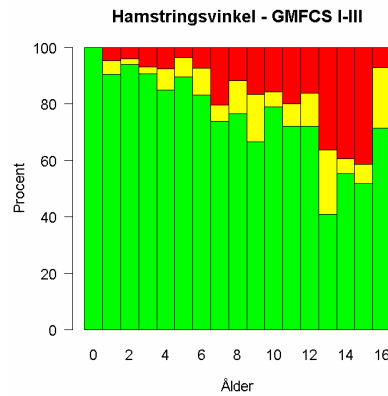
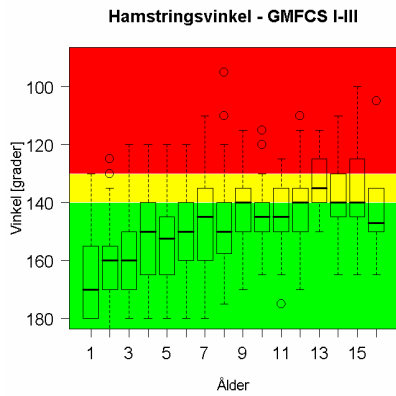
### Hip abduction



### Hip extension

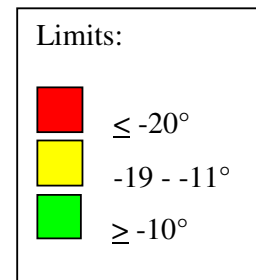
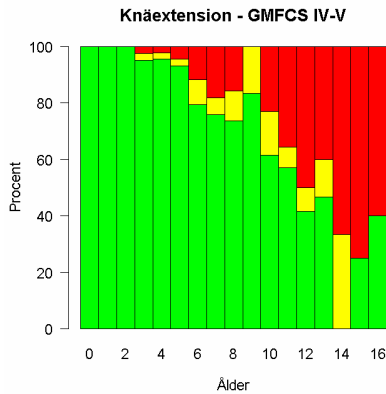
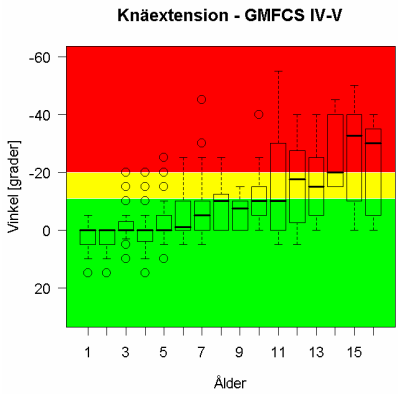
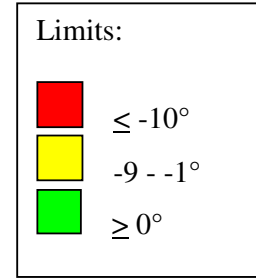
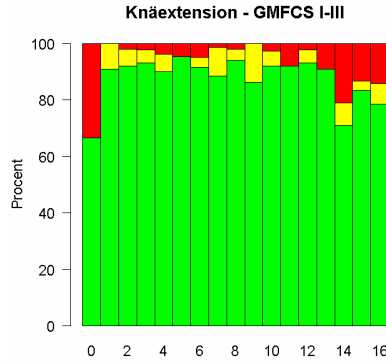
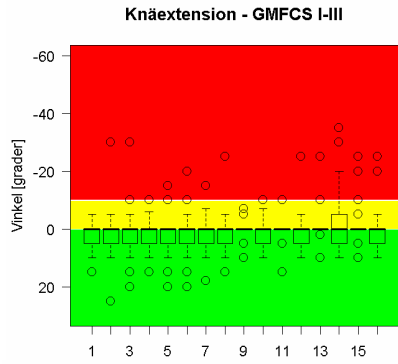


### Hamstring Angle

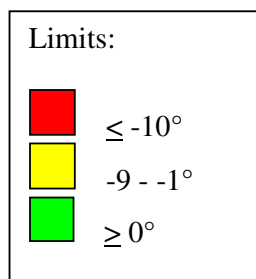
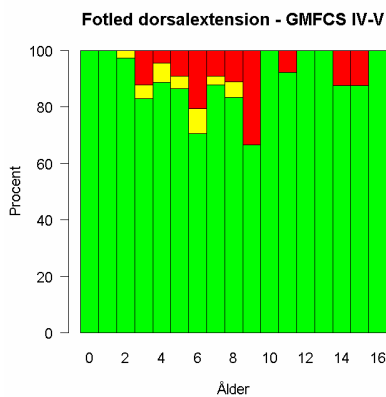
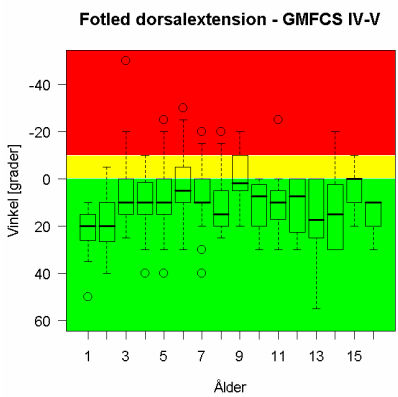
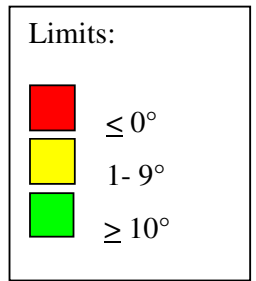
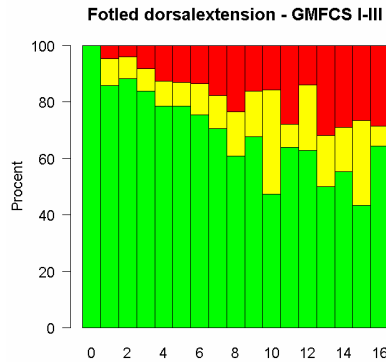
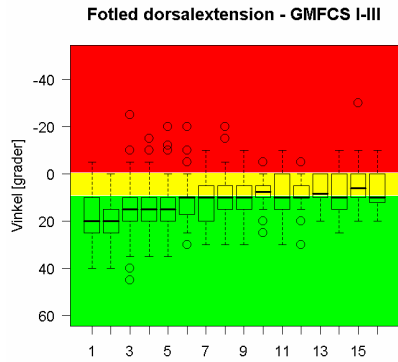




**Knee extension**



**Ankle joint – dorsal extension with extended knee**



### *Commentary range of motion- lower extremities*

The results concerning the lower extremities consist of examination data on 1335 children, aged 0-16 years of age (children not evaluated according to the GMFCS are excluded). The total number of reported measurements for hip, knee and feet are illustrated in the box plots. The figures to the right of the box plots illustrate the distribution in percent (%) of the number of passive range of motion measurements for each age group in relation to green, yellow or red limits. As of this year, with these figures, we are happy to present measurement values on children under the age of one. It is a positive development that children are detected and monitored in CPUP even in their first year, but as there are still few measurements in the lower and higher age groups, the results should be interpreted with caution.

*The hip abduction* measurements on children and adolescents, who are being monitored regularly in CPUP, shows that about 70% of the values (n=1112) were all normal. The yellow measurement values, which should be controlled /treated were about 10% and the red, clearly pathological values about 20%. This is representative for the children at both the GMFCS I-III and IV-V levels. The red values are represented in all age groups except for children under the age of one.

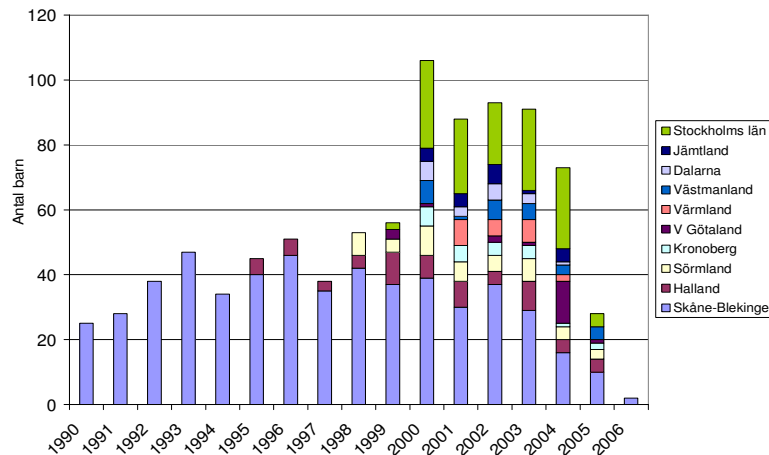
Decreased passive *hip extension* seldom occurred. 95% at GMFCS levels I-III and 84% at GMFCS levels IV-V out of a total of 1083 measurements, showed a green limit value. Yellow limit values for GMFCS I-III were not reported. The red limit values were represented at both GMFCS I-III and IV-V levels and in both the higher and lower age groups.

The measurement of the *hamstring angle* (n=1129) shows that about 75% of the values fall within the green limit value. Within GMFCS I-III, the number of yellow and red limit values represents about 20% of measurements. The corresponding number at GMFCS levels IV-V is 26%. The red limit values mostly occur in the school starting years, ages 6-7, as well as in the adolescent years and upwards. At GMFCS level I-III, 10% of the *knee extension values* were yellow or red. For GMFCS IV-V the corresponding value was 20%. The clearly pathological values already occurred at the age of three and became more common with progressing age. At GMFCS levels I-III, yellow and red limit values were found in 30% of the measurements of the *dorsal extension of the foot* in all age groups. For the group at GMFCS levels IV-V, 85% of the measurement values of the dorsal extension of the foot were found within the green limit value.

We can ascertain that we noted deterioration in range of motion ability in several joints of the older children irrespective of the severity grade of the functional impairment. We can also see that the children with a more pronounced functional impairment (GMFCS IV-V) have a tendency to a higher degree of deterioration with progressing age. For the children and adolescents, whose gross motor function ability were classified at GMFCS level I-III, deterioration in hip abduction, hamstrings angle and dorsal extension of the ankle joint was noted in the higher age groups. They also had a marginal deterioration of hip and knee extension respectively. The possible reason for this could be that the children have a decreased mobility with progressing age and/or that the children born in the 1990's have received a less effective treatment than the children born later. We do not know as yet, how the continued development of range of motion status up to adulthood will develop. After the age of 15, the rapid growth phase of the skeleton has ceased, which should diminish the risk of deterioration. Possibly, more active contracture prevention during the pubertal growth spurt period is important to obtain a permanent improvement in mobility. It could also be discussed whether children in the teenage years should be examined more than once a year in CPUP.

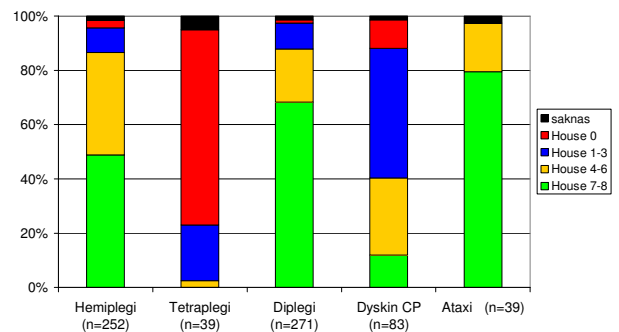
## Hand and Upper Extremity Function

*The number of children from respective region, who have been registered for arm and upper extremity function in relation to year of birth (n= 887).*

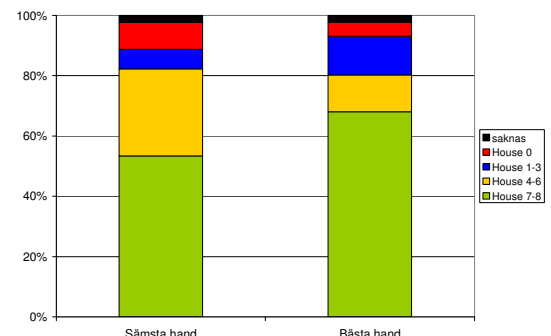


In 2006, 887 children and adolescents (0-16 years of age) were registered with regards to their arm and upper extremity function in the Occupational Therapist assessment form in CPUP, 535 (60%) were from the regions of Skåne and Blekinge, where reporting of arm and upper extremity function started in 2002. The new regions, which have been added gradually have often commenced with the younger children. Data concerning the older age groups therefore mainly comes from the Skåne and Blekinge area. Stockholm has partly used another assessment form and unfortunately, we can therefore not account for certain data on these 115 children. With the introduction of 3C during 2007, information from all the participating regions should be complete.

Similar to last year, an equally large number of children with hemiplegia and diplegia (45%) had “mild” hand problems (MACS I). For the children with hemiplegia, almost 30 % were reported as MACS II, which among other things mean they do not have full bimanual function but often use different strategies to be able to carry out their daily activities. In children with spastic diplegia and ataxia, all MACS levels were represented and 65% were independent in their daily manual activities (MACS I + II). All children with tetraplegia and 85% of children with dyskinetic CP had very reduced or lacked hand function completely (MACS IV-V).

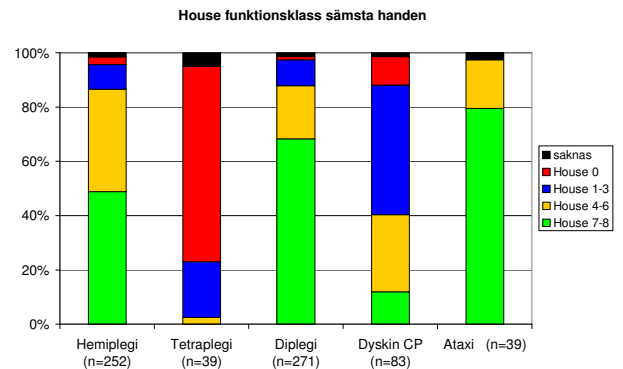


Hand function according to the House Classification is evaluated for each hand separately on a scale between 0 and 8. The four main groups are accounted for here, a hand which is used spontaneously and independently from the other hand (7-8), an active assisting hand (4-6), a passive assisting hand (1-3) and a hand not used at all (0). Almost 50% had two hands, which were classified at the highest functional level. 25% had an active assisting hand on their most affected side. Amongst the 704 children who

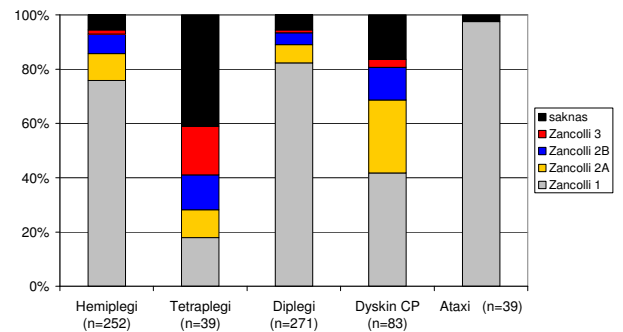


had been classified, 5% did not use any hand at all in their activities.

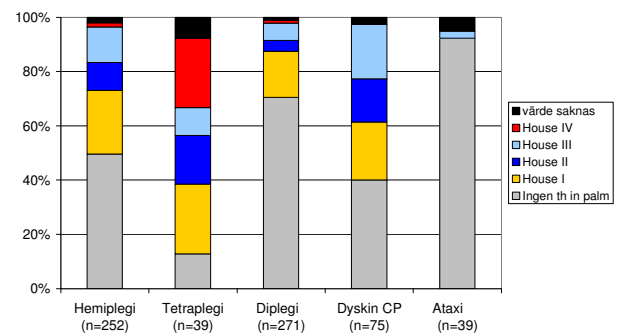
The function in the most affected hand was classified lower in hemiplegia than in diplegia. If one compares House functional classifications with the distribution of MACS levels (see figure above), there is a clear difference concerning the children with dyskinetic CP. Many of these children seem to have a rather good grasping ability in their hands, yet they have pronounced problems in their activities of daily living, 85% were classified at MACS level IV-V. Maybe, lacking voluntary gross motor function control in shoulders and elbows could explain this difference? In the future, one might in CPUP consider including some measurement of the ability to place the hands adequately. One can also speculate if further treatment input is needed for this group.



The Zancolli classification is used to determine the degree of muscle tone in the wrist and finger flexor muscles. The classification is done by asking the child to actively extend the wrist and fingers simultaneously. At grade 1, tone is normal or only slightly increased, at grade 3, there is a complete inability to extend the wrist and fingers. It can be difficult to assess a child who is unable to co-operate and therefore values are sometimes missing in children with tetraplegia and dyskinetic CP. 80% of all the reported children were evaluated as Zancolli 1, which coincides rather well with the fact that no tightness was reported at passive wrist extension with straight fingers in 88%. All the children with ataxia could actively extend their wrists and fingers.



41% of the children were reported to have problems with thumb-in-palm, in at least one hand. However, the assessment is difficult and one can suspect a certain over-diagnosis. The assessment should take place when the child actively opens his/her hand to grasp. An inactive, relaxed hand can appear to have an adducted thumb as with House type I without any existing spasticity. Half of the children with hemiplegia had thumb-in-palm, but almost no children with ataxia. Type IV where all muscles are spastic only existed in children with tetraplegia.

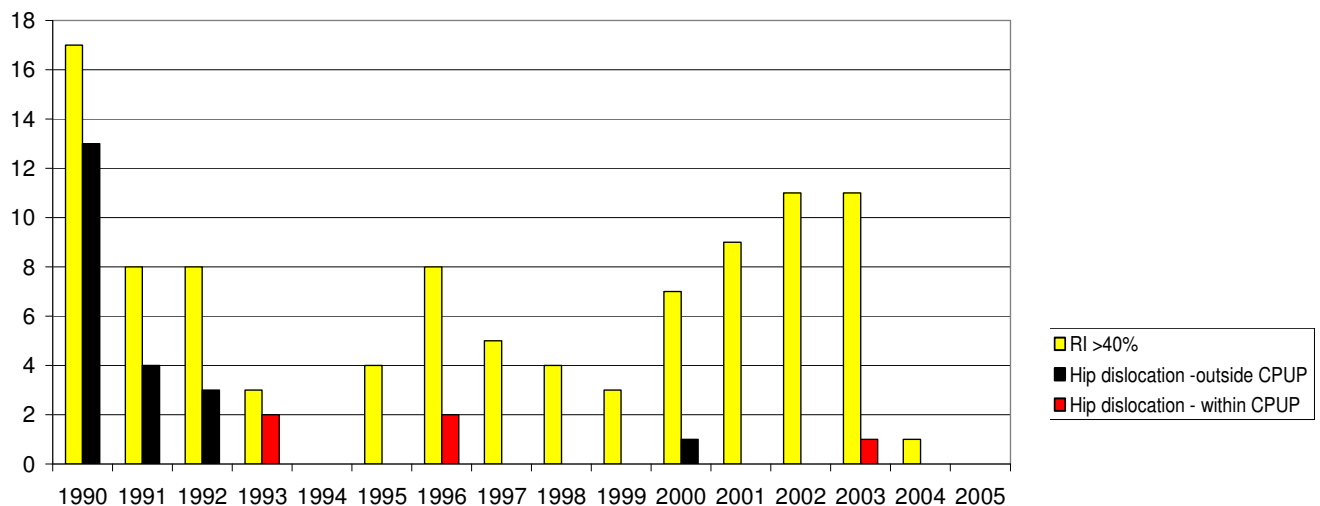


## Hip dislocation

The goal with hip screening is to prevent hip dislocation, i.e. hips with a Reimer's Index (RI) 100%. We know through an analysis of the screening results in Skåne and Blekinge that hips with an RI >40% often need surgical intervention in order not to deteriorate and develop hip dislocation. Hips with a RI between 33% and 40% need to be observed closely and sometimes surgical intervention is needed.

We have hip x-ray results for 1067 children. According to the new health care programme (please see [www.cpup.se](http://www.cpup.se) for more information) children at GMFCS level I do not need to be radiologically examined. One county council (Sörmland) has not reported any hip results for 2006. We have chosen to present hips with an RI >40% and hips with an RI of 100%, i.e. dislocated hips (see figure).

Proportion of children with RI >40% and hip dislocation related to year of birth



*The proportion of children (%) with a Reimer's Index >40% and the proportion of children with a hip dislocation (RI 100%) in relation to year of birth. The children born in 1990-1991 act as a reference group for Skåne and Blekinge and have not participated in the hip prevention programme. The red bars showing hip dislocations in children born after 1991 represent 5 children in total.*

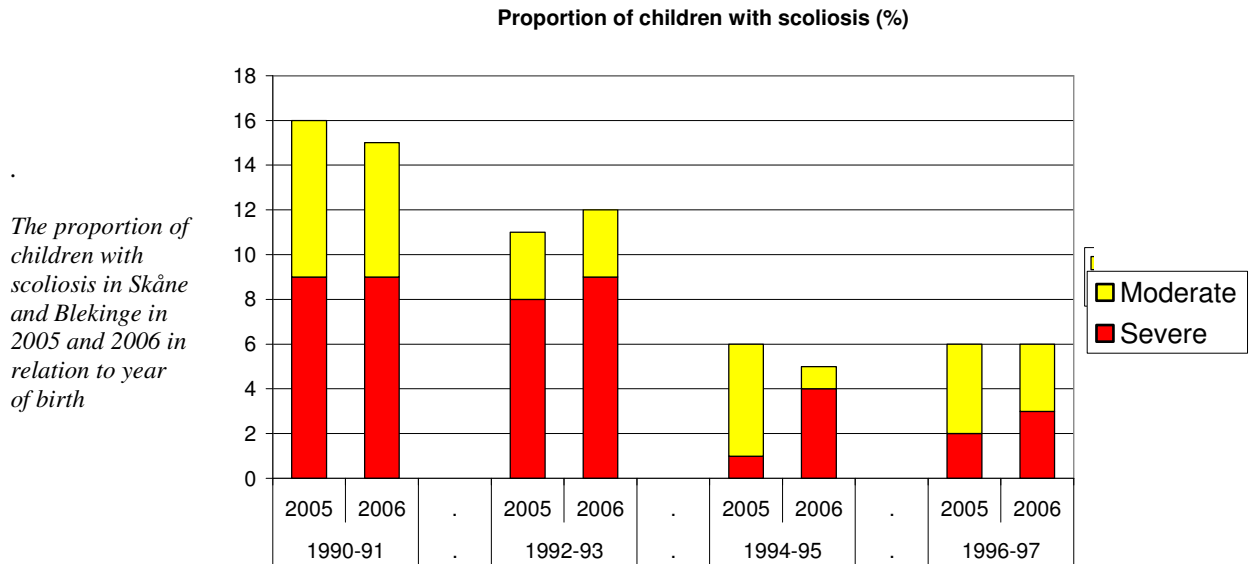
We know that 15-20% of the children would have developed hip dislocation if no preventive measures had been taken. The children born in 1990-1991 act as the reference group in Skåne and Blekinge, i.e. they have not participated in the hip screening programme. The same encompasses a child born in 2000 from another region. Apart from these, there are four children who have been reported with hip dislocation. Three children (red bars) have been deemed to be in such bad general health that preventive surgery could not be performed.

One child born in 1992 with hip dislocation had moved in from a county that, at the time, did not participate in the CPUP programme. So far, no hip dislocation has occurred due to late entering in the programme.

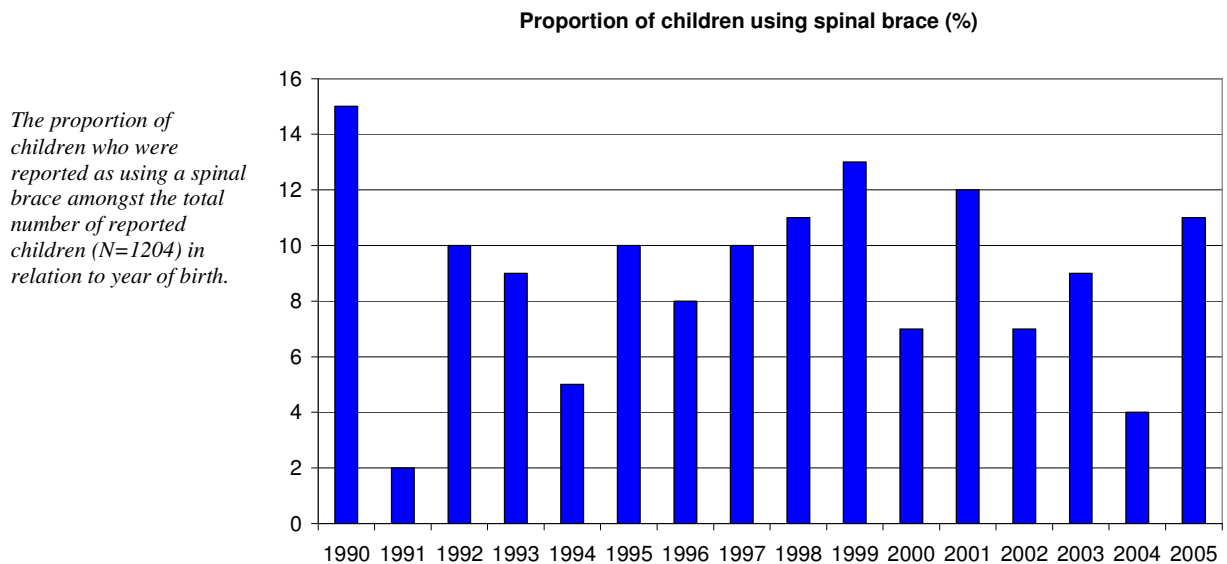
The results indicate that prevention of hip dislocation in CPUP continues to be successful. It is too early, as yet, to analyse the results of different preventive measures /operations.

## Spine - Scoliosis

The intention with the spinal follow-up is to try to decrease scoliosis that would need surgical intervention and to discover this type of scoliosis in time. We have chosen to present the proportion of children with scoliosis that was considered to be moderate or pronounced. As scoliosis often is likely to develop in the adolescent years, the results so far only represent the counties of Skåne and Blekinge. The decrease in the proportion of children with scoliosis can partly be explained by differences in age, but even after correction for age, a decreasing number of children with scoliosis can be seen, even in the higher age groups.

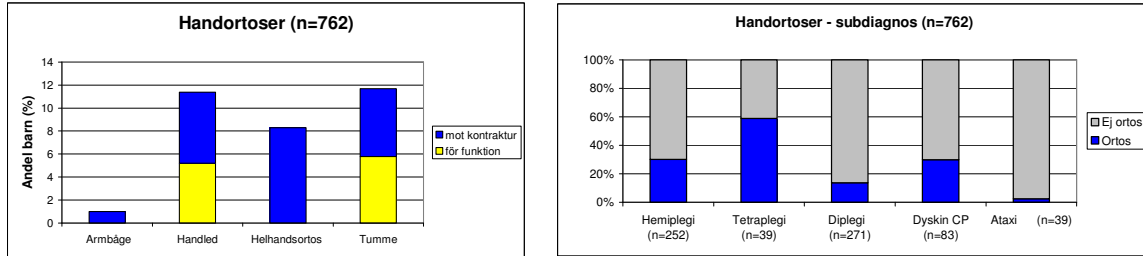


Nine percent of the total number of children reported wore a spinal brace. The distribution in relation to age (year of birth) is shown in the figure below. In Skåne and Blekinge, we have seen great variation in the use of spinal brace between different habilitation districts, but it is still too early to analyse differences between the various regions. The most likely reason for the high number of young people born in 1990 using a spinal brace is that there are many patients suffering from scoliosis and windswept position in that particular age group.



## Use of Orthoses

### Orthoses upper extremities

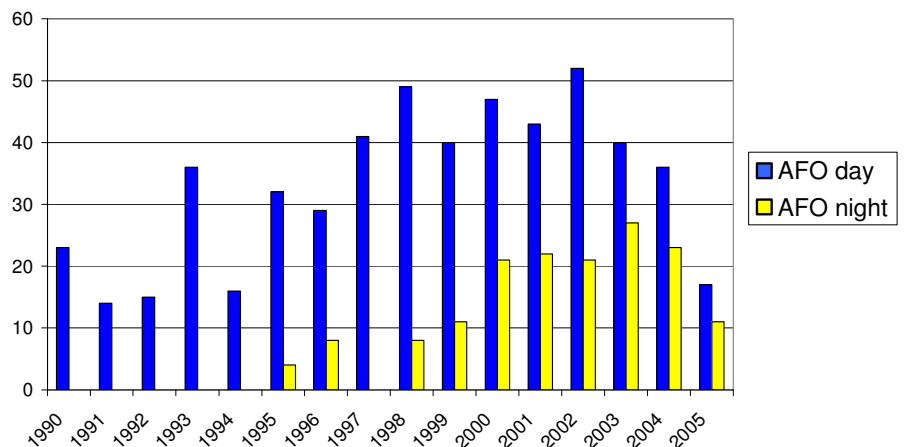


Some type of hand orthosis was used on a regular basis by 24% of the children. Wrist and thumb orthoses were almost equally common and often used to facilitate function as well as for contracture prevention. Hand orthosis was used only as contracture prophylactic. Elbow orthoses were more uncommon. There was a great variation in the use of orthoses between the different CP subtypes. Only one child with ataxia as opposed to 60% of children with tetraplegia used an orthosis. With the introduction of 3C in January 2007 the actual time wearing the orthosis (more or less than 6 hrs/day) will be registered.

### Orthoses lower extremities

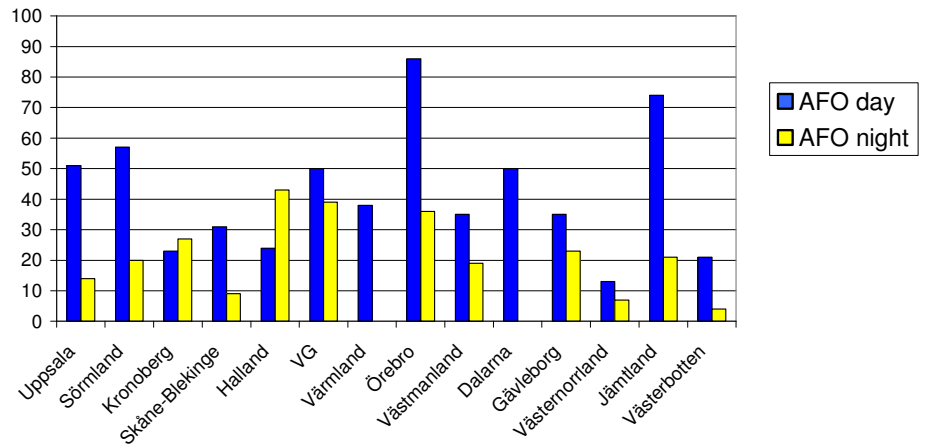
The most common type of orthosis is AFO's (ankle-foot orthoses) and standing frames/standing shells. AFO's are used in the daytime by 38% of the children and by 14% of the children at night. The distribution related to year of birth is shown in the figure below. We can see that AFO's during the day were used in all ages with a slight decrease in the adolescent years. AFO's at night had a more clear age profile with an increase up to about 3 years of age and a decrease thereafter.

*The proportion of children (%) who used AFO's at day or night amongst the total number of reported children (N=1204)*



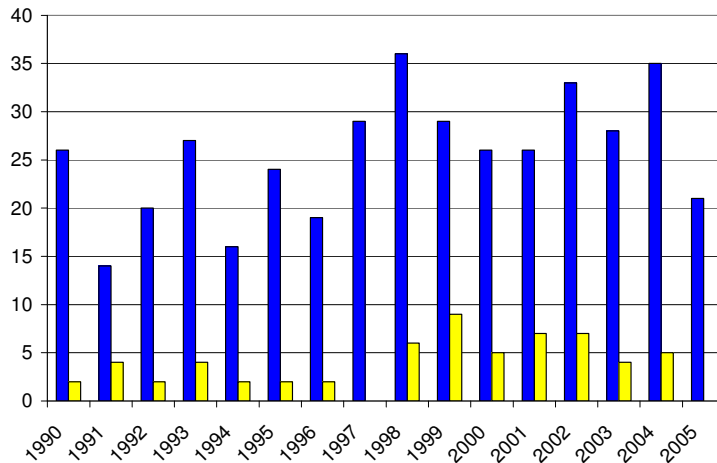
We can see a great variation in the use of AFO's between the various regions. The differences are so great that, despite of the fact that relatively few children are reported as yet, some differences are statistically significant.

*The proportion of children (%) using AFO's at day or night amongst the total number of reported children born in 2000-2003 (N=660) distributed according to region*

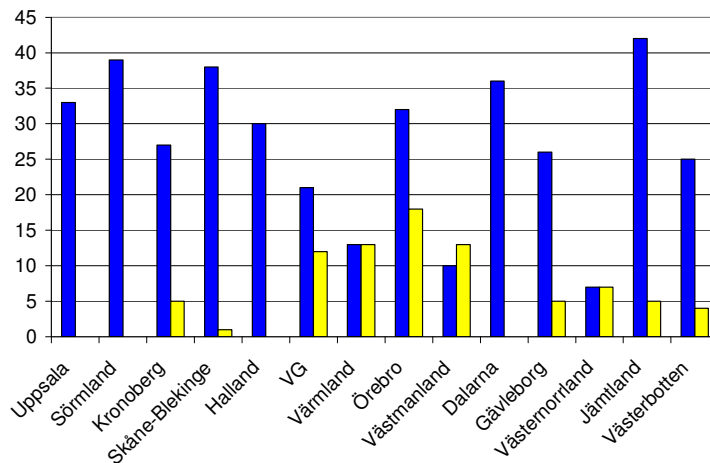


Standing frame/standing shell (standing in a shaped orthosis reaching up to waistline or higher) was used by 27% of all the reported children. Apart from that, 5% were using other types of standing support (Easystand, Tilt table, Gazelle etc). The use of standing frame/standing shell does not have the same age profile as AFO's, 15-20% use it, even in the adolescent years. In the counties of Västra Götaland, Värmland, Örebro and Västmanland, it is foremost Tilt table that was used "as other standing support".

*The proportion of children (%) using standing shell (blue) or other standing support (yellow) amongst the total number of reported children (N=1204).*



*The proportion of children (%) using standing shell (blue) or other standing support (yellow) amongst the total number of reported children born in 2000-2003 (N=660) distributed after region*



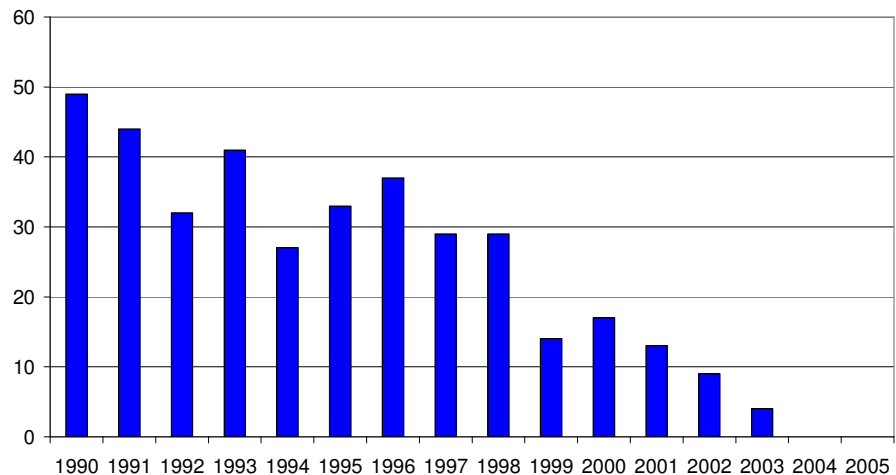


## Operations

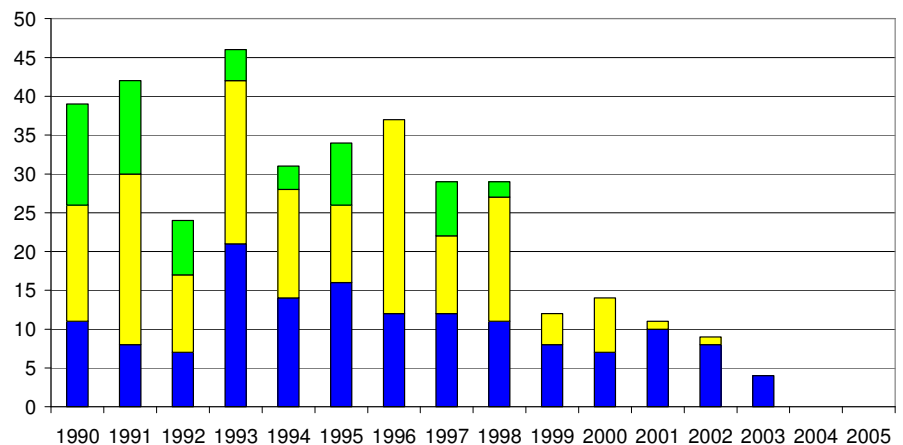
Out of a total of 1084 children (five regions have not reported operations yet) 223 have been reported as having had one or more surgical procedures performed. The proportion of children who have undergone a surgical procedure in relation to year of birth can be seen in the figure below. Out of the 223 children who have undergone surgery, 96 have had preventive orthopaedic surgery against hip dislocation (adductor-psoastenotomy, femoral osteotomy, pelvic osteotomy). Achilles tendon lengthening (or gastrocnemius lengthening) have been performed on 89 children. The third most common type of surgical intervention is subtalar arthrodesis, which has been performed on 27 children. Only 9 children (1%) were reported as having undergone hand surgery in the last year. It should be noted that children under the age of six made up the basis for half of the material concerning the upper extremities.

It is still not possible to do a closer analysis of the distribution of operations performed between the different regions. There is, however, a large spread in the frequency of operations due to the occurrence of contractures between different districts and regions. In the future, it will be interesting to analyse these differences in relation to possible differences in range of motion status and function.

*The proportion of children (%) amongst the total number of reported children (N=1084), who underwent orthopaedic surgery at some stage up until 2006 in relation to year of birth.*



*The proportion of children (%) amongst the total number of reported children (N=1084), who underwent surgery for prevention of hip dislocation, achilles tendon lengthening or subtalar arthrodesis, at some stage up until 2006 in relation to year of birth.*

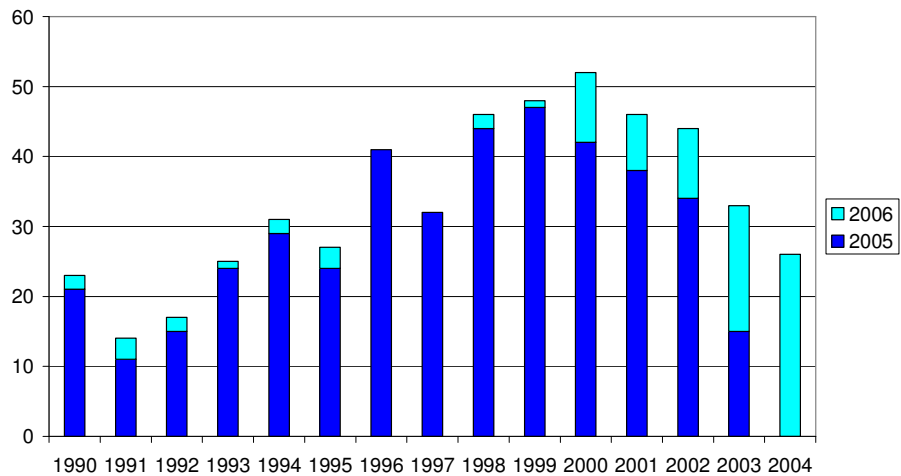


■ Hip - prevention ■ Achilles lengthening ■ Subtalar arthrodesis

## Spasticity reducing treatments

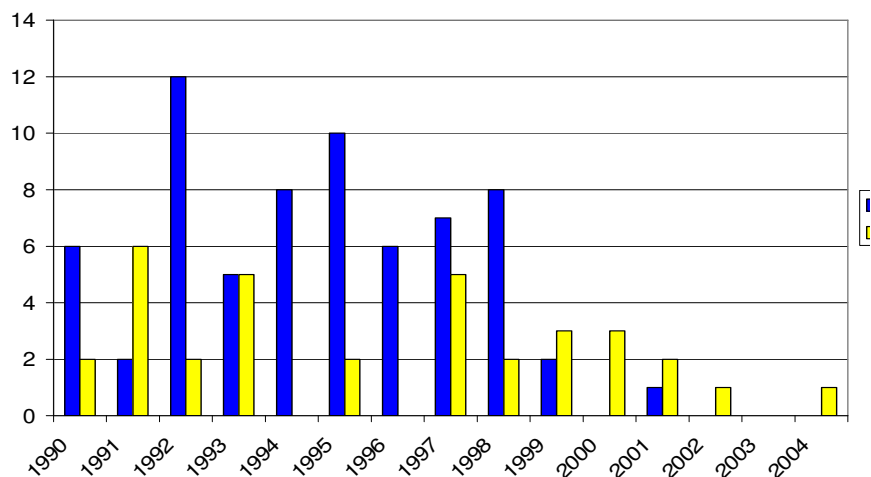
The proportion of children, who were reported as having had botulinum toxin treatment at some stage are shown in the figure below. The material is still too limited for an analysis to be made of the geographical differences or the treatment results. All regions have, however, reported children who have undergone treatment with botulinum toxin. The most common muscle group that is treated is the gastrocnemius-soleus muscles. We can see that the proportion of treated children, have increased since 2005. Botulinum toxin treatment in the arm and hand had been given to 56 out of the 887 children (6.3%) who were reported in the upper extremity protocol. Out of these, 33 had hemiplegia, 12 diplegia, 3 dystonia, 4 tetraplegia and 4 had an unclear CP type. All MACS levels were represented.

*The proportion of children (%) amongst the total number of reported children (N=1204), who had been treated with botulinum toxin, in relation to year of birth. The number of recently added children, who have been treated in 2006 is shown in light-blue colour.*



The proportion of children who had been treated with Baclofen pump or had a selective dorsal rhizotomy operation (SDR) performed is shown in the figure below. As a rhizotomy often is performed in the 3-6 year age group, the figures show a decreasing trend in later years. As baclofen pump is inserted and used in all age groups, the lower number in the younger children is expected. It is still not possible to determine the development in the frequency of baclofen pump treatment.

*The proportion of children (%) amongst the total number of reported children (N=1374) who had been treated with either rhizotomy (blue) or baclofen pump (yellow) in relation to year of birth.*



## Summary

As of January 2007, all county councils/ regions in Sweden are now participating in the CPUP programme. In Norway, Helse Sør and Öst are participating, corresponding to half the population of Norway. This report presents the situation in 2006, when regions corresponding to 85% of the population in Sweden had joined the programme. In the age groups where all the participating regions are reporting, the number of children in the register corresponds to a prevalence of 2.0/1000, which indicates that most children with CP in the present age groups participate in CPUP.

All participating regions have not yet reported in all parts of the CPUP programme. However, we can see that more regions gradually are starting to report in all assessment forms. The assessment form for the neuropaediatricians has been drawn up during 2006 and reporting started in 2007. Our hope is that we will soon reach the time when all regions are registering in all assessment forms. This is necessary in order to get a complete picture, both over the individual child and for CPUP as a quality register.

One of the goals of CPUP is to prevent hip dislocation. So far, it seems as if we are able to achieve this in the entire country. Let us not forget that before the CPUP programme started, every 10<sup>th</sup> child with CP developed hip dislocation, so it is a major improvement that has been achieved. We now know, that many hips already start to lateralize around the age 2-4. Therefore, it is important that we continue to be active in identifying all children with suspected CP as early as possible and to monitor them in CPUP to enable early intervention.

We are now starting to see the differences in treatment routines between the various county councils/ regions. For example, there is great variation in the use of orthoses. One way of evaluating different treatment methods is to point out differences in treatment, and then analyze the reason for the differences and if there is any difference in the treatment results. For example, do county councils where a vast number of children use AFO's or standing frames show a better result than in the regions where fewer children use AFO's? We do not know, as yet, whether over or under treatment takes place.

The new spasticity reducing treatments are also important to evaluate. We can see an increasing number of children who are being treated with botulinum toxin. At the same time, we can see a decreasing number of children who are having a rhizotomy performed. Hopefully, within a few years, the long-term results of these new methods can be analysed.

We can see that the joint mobility is poor in the older children. The deterioration is more pronounced in children at MACS levels IV-V and GMFCS levels IV-V. In the future, we hope to see how different treatment alternatives can affect the development of joint mobility. For example, does the increased use of botulinum toxin after 1998 mean that the joint mobility will improve for the children born in later years? How can different routines in the use of orthoses affect the joint mobility development?

We can also see that problems with hand function are common in cerebral palsy and that the problem exists within all the CP subtypes. Therefore, it is necessary to monitor *all* children with CP in terms of hand function. It is too early to determine how successful we are in our preventive work, but CPUP have already meant great qualitative gain by increased focus on the upper extremities and a better comprehensive view on the child's range of motion ability.

Ever since the start of CPUP, an increasing number of scientific articles have been written and we now have a vast material, which is increasing in size and this provides us with a unique opportunity to evaluate and analyse different aspects of CP. The possibility of using informa-

tion from the CPUP database also increases from year to year, and we hope that as many people as possible will be interested in taking advantage of this.

## Goals for 2007- Evaluation

We will, in co-operation with NKO during 2006, be preparing for netbased reporting of information to the CPUP database, which will be done by transferring the follow-up assessment forms to Comporto, an internet based reporting function. Test drive is planned for autumn 2006, and the plan is to start using Comporto from 2007.

*Commentary: Completed*

The regions, who are not yet participating in CPUP, have all advertised that they have plans for and wish to participate in the programme. This means that the entire country should participate in CPUP from 2007.

*Commentary: Completed*

Continued development of CPUP/CPOP is taking place in Norway. We hope that Denmark will start to participate during 2007.

*Commentary: Continued development and co-operation with Norway. Discussion and evaluation for possible participation in 3C continues. Co-operation with Denmark has not developed during the year.*

Education in the CP classifications is continuing in connection with the CPUP days on the 21<sup>st</sup> -22<sup>nd</sup> of September 2006. An educational CD will be presented and has been translated into Swedish for CPUP in connection with the creators within SCPE (Surveillance of Cerebral Palsy in Europe) and with financial support from the Folke Bernadotte Foundation.

*Commentary: Completed*

During 2006, work groups have made propositions to revise the assessment forms, which are used by the physiotherapists and occupational therapists. The propositions should be submitted for comments and new versions are planned to be in use as of 2007.

*Commentary: Completed*

In consultation with the reference group in neuropaediatrics, a work-group have constructed a assessment form for the reporting of for example CP-subdiagnosis, other impairment/disorders and growth etc. The proposal has been submitted for comments and is planned to be in use as of 2007.

*Commentary: Completed*

PM for hip screening should be revised based on the experiences we have had so far. The proposal is to be submitted for comments and is planned to be in use as of 2007.

*Commentary: Completed*

## Goals for 2008

Contacts have been made with representatives for the Habilitation Services for Adults in Sweden. An investigation has been initiated in order to map out the prerequisites for the continued follow-up for adults with CP and what form this follow-up should take. This investigation should be completed during 2008.

The contents of the different assessment forms in CPUP should be optimized, so that we, as clearly and easily as possible, can receive the information we need in CPUP, both to follow the individual child and also for CPUP as a National Quality Register.

Continued development of and education in the 3C system. The system should be as user friendly as possible. Different ways of being able to trace technical faults easily and different ways of tracking children with signs of deterioration will be worked out.

Development of the Norwegian version of 3C, and continued efforts to be made for Norway to be able to report in 3C. The goal is that routines for reporting in 3C from Norway will be established in 2008.

Translation of parental information to the foreign languages that are needed.

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