



# European working group on psychosocial aspects of children with chronic renal failure

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## Family education program chronic renal failure – concept of a psychological and educational intervention for children and adolescents with chronic renal failure and their families

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Introduction: Chronic renal failure (CRF) in children and adolescents is a burden and challenge for the affected patients and their families. CRF impairs health related quality of life (HRQoL) and psychosocial adjustment in all members of these families. Furthermore, a high degree of responsibility and motivation of the child and its relatives is essential to achieve treatment goals. Sustained motivation for therapy in the long-run is hindered, because effects of medication are often not visible for the patient and his family in CRF e.g. delay of progression (antihypertensive treatment) or prevention of secondary effects (secondary hyperparathyroidism, cardiovascular risk).

Family education programs mean interventions with medical and psychological aspects. Instead of briefing or instructing these education programs are multidisciplinary with several professions and are performed in a group. Around 5-7 children of similar age with CRF and their parents as well as siblings attend to the program. The basic concept is empowerment of the family members.

For common chronic diseases in childhood (e.g. asthma, diabetes,...) these education programs are components of the integral therapy and part of disease management programs. In rare diseases these education programs are arising in meanwhile: e.g. we developed a program for children with nephrotic syndrome and their families in 2007. For children with CRF and their families, however, a specific education program does not exist.

Methods: We developed a modular multidisciplinary education program for children and adolescents suffering from CRF and their families. Modularization was performed in accordance to the concept of ModuS (modular patient education program for children and adolescents) of the competence network of education programs in children and Adolescents (Kompetenznetz Patientenschulung im Kindes- und Jugendalter) founded by the ministry of health (BMG AZ IIA5-2509KI006/314-123006/04).

The main characteristics of the program: This concept is based on the dichotomization into generic and disease specific modules. Generic modules contain disease independent topics like coping, education of chronically ill children and their siblings,... CRF specific modules covers physiology and pathophysiology of the kidney (e.g. arterial hypertension) as well as treatment options which are taught according to the cognitive state of the children and adolescents with different modules for every age group. Nutrition in CRF is also a big issue. Children and parents are trained separately to allow age-based teaching and to consider the different expectations. This two days lasting program is planned to accompany the patients regularly until transfer into an adult care. The team is formed of nurses, doctors, psychologists, pedagogues and dieticians. Evaluation of the programs is performed by qualitative methods (structured interviews), standardized questionnaires (HRQoL, psychosocial adjustment) and medical parameters.

Conclusion: The family education program chronic renal failure is the first multidisciplinary and modular education program for children with CRF and their families and is thought to help the affected persons to experience themselves as autonomous members in the health-team (empowerment). This program could play a role within an integrated concept of treatment for children with CRF and should be financed by the national health system.

## The Teaching Parents Study (Phase 1): a pan-Britain, qualitative exploration of multi-disciplinary teams teaching parents to deliver home-based care of childhood chronic kidney disease

Swallow V, Allen D, Williams J, Smith T, Crosier J, Lambert H, Quizlbach L, Wirz L, Webb NJW, Nightingale R

**Background:** Research on long-term childhood condition management often overlooks the complexity of health care professionals' roles; as well as taking overall responsibility for the child's condition; professionals often hand over day-to-day responsibility for clinical care to parents. To support parents in their clinical care-giving role, multi-disciplinary teams caring for children with chronic kidney disease, therefore, spend considerable time teaching parents to safely manage their child's condition at home. However, the strategies multidisciplinary team members use to educate parents from early in the renal journey have received little research attention. The aim of phase 1 was, therefore, to explore professionals' accounts of the parent-educative aspect of their role.

**Methods:** As this was a previously under-explored issue, a qualitative, inductive design was used involving individual and focussed interviews. Multi disciplinary team members in the 12 children's kidney units in Britain were invited to discuss the parent-educative aspect of their role. Written consent was obtained from all participants. Data were collected through 13 focus group and 7 individual research interviews. A total of 112 professionals (7 clinical psychologists, 9 dieticians, 30 doctors, 48 nurses, 3 pharmacists, 7 play workers, 6 social workers, and 2 therapists) participated. Interviews were digitally recorded and later transcribed. Data were anonymised, and analysed using Framework Technique.

**Results:** Three themes emerged.

1. Initiating knowledge and skill development
2. Promoting and monitoring knowledge and skill development
3. Multi-disciplinary team working

This presentation focuses on Theme 3 and specifically the sub-themes:

- Role allocation & differentiation
- Sharing information within the team

Participants used the term 'we' widely, this could denote both the collective identity when they referred to the intradisciplinary 'we' (for e.g. 'we' as a local or national group of doctors, nurses, dieticians, play workers), or the individual identity when they referred to the interdisciplinary 'we' (ie as the multidisciplinary team). Examples of the many quotations from the eight different disciplines represented in the data will be presented to illustrate these sub-themes.

Discussion and conclusions: The concept of Distributed Expertise helps explain how multidisciplinary teams teach parents, it captures the idea that different professionals offer different kinds of expertise to the co-management of a child's condition; this includes specialist knowledge and the resources to promote that knowledge. Distributed expertise also explains the team's capacity (rather than just the individual's) to learn, act on and transform the problems of practice when supporting parents to manage their child's condition. Communication within and between disciplines is essential to promote parents' safe and effective home-based clinical care giving. At different points of the trajectory the key professional teaching the parents may vary. Whilst recognising the team's capacity to support parents as they learn to administer clinical care to their child it is also important to recognise each discipline's distinctive contribution. These findings informed data collection in Phase 2 of the study, which aimed to obtain a detailed understanding of parent-professional interactions during teaching/learning encounters in the management of childhood CKD.

Funder: Kids Kidney Research

## The Teaching Parents Study (Phase 2): Longitudinal, observational research around how parents learn to deliver home-based care of chronic kidney disease

Swallow V, Allen D, Williams J, Smith T, Crosier J, Lambert H, Quizlbach L, Wirz L, Webb NJW, Nightingale R

**Background:** In the management of childhood chronic kidney disease the best possible clinical care-giving is essential to minimise kidney deterioration and optimise children's well-being. Parents often perform the majority of clinical care-giving at home so to promote and support their care-giving it is important to understand parents' views about and experiences of learning to deliver their child's care. However, few data exist which explains how parents learn to deliver home-based care of long-term childhood conditions. This paper reports on phase 2 of The Teaching Parents Study; the aim was to obtain a detailed understanding of parent-professional interactions during planned and ad-hoc teaching/learning encounters in the management of childhood chronic kidney disease.

**Methods:** As this was a previously under-explored area of clinical practice, a longitudinal, observational study was undertaken. In accordance with the principles of ethnographic research, a Children's Research Practitioner spent an extended period of time undertaking six focussed case-studies in two UK children's kidney units. Parents of six recently referred children were recruited using a purposive sampling approach. To achieve maximum sampling variation children were selected according to gender, ethnicity, age and the type of clinical care-giving needed. Snowball sampling was used to identify multidisciplinary team members involved in managing the child and in teaching and supporting parents; these professionals were also invited to participate.

Data gathering lasted six months for each case-study and involved a combination of: minimally-obtrusive observations of professional-parent interactions in the clinical setting and families' homes; de-briefing interviews to obtain individuals' views on selected interactions; focussed 'verbatim' field-notes, and case-note reviews. The focus was on communication between parents and professionals (plus the intervening researcher if appropriate) as parents learned the clinical care-giving skills and information needed. Dialogue during observations and interviews was digitally recorded, transcribed and anonymised. Participant' initial written consent was re-affirmed verbally as data collection

proceeded. Data were managed and analysed using an iterative-inductive approach supported by Framework.

Findings: Four main themes were identified:

1. Teaching & Learning Processes;
2. Facilitators to Teaching & Learning;
3. Barriers to Teaching & Learning;
4. Ambivalence within Teaching & Learning Encounters.

In this presentation, excerpts from the researchers' field-note observations, and parents' and professional' qualitative accounts, will be used to illustrate and discuss these themes.

Conclusions: This detailed insight into the social context in which parents learn to deliver clinical care to their child addresses an important gap in understanding this complex intervention. The findings will help to inform professionals' care delivery, educational curricula, and health-service policy and may also be valuable to professionals teaching parents caring for children with other long-term conditions. This study forms part of a phased approach to developing and evaluating a complex intervention that addresses parents' learning needs in care delivery.

Funder: Kids Kidney Research

## Adolescence – Non-Adherence –Transition: Interdisciplinary Challenges in Organ Transplantation

Petersen I, Hamburg

Life-long medication adherence is a critical component of health care management for transplant recipients. Non-adherence is the main cause for organ rejection and graft loss - especially in adolescence and young adulthood. Adolescence is an extreme vulnerable period with several developmental tasks like peer-group acceptance or detachment from parents. Changes and destabilization take place in all life domains. Biologically, this is reflected by extensive brain development. The varying pace of development in different brain areas leads to an imbalance between brain structures which influence behavior. As a consequence, emotions and the "rewarding system" of the brain dominate the power of judgment and action planning. Adolescent behavior is characterised by extreme changes, inconsistencies and unstable cognitive abilities. Risk behavior and impulsivity is increased, action planning and self-control is reduced.

Furthermore, adolescent transplant recipients show a strong inclination to be and to behave just like other people of their age. Although they are aware of their chronic illness and their doctors' recommendations the pressure towards health risk behavior might be stronger. All these factors present unfavorable conditions for adherence. Therefore, it is not surprising that the prevalence of non-adherence in adolescent transplant recipients is significantly increased (30-53%) compared to children (3-19%). A number of causes for non-adherence are discussed. However, the role of the parents is often underestimated and not well understood. Parents have a strong influence on their child's development and behavior. According to research findings parenting stress is significantly related to poor adherence. Furthermore, parental adherence is a role model for the young transplant recipients. However, parental beliefs regarding immunosuppression are often negative and many parents hope that immune tolerance will be achieved in the near future.

Studies show that adherence in pediatric transplant recipients further decreases when they transfer to the adult clinic. Recently, several studies and interventions have been initiated worldwide with the aim to improve the adherence of adolescents. Meanwhile, there is an increasing recognition that adolescent patients who received a transplant at a young age represent a group with special requirements, particularly with regard to the critical

transition phase. In this context "transition" means a multidimensional, active process which should be a purposeful, planned movement of transplant recipients from pediatric to adult-centered healthcare systems. In contrast "transfer" means just the change in the location where care is provided on a specified date. Transition includes two parallel processes: 1. the transition of responsibility for health management tasks from the parents to the patient and 2. training programs in the pediatric clinic and preparation for the transfer to the adult clinic. Transition can be divided into three phases: the early transition (10-12 yr), the middle transition (13-15 yr) and the late transition (• 16 yr), with special tasks in each phase.

Responding to the particular needs of adolescent transplant recipients several transition programs and transition clinics have been established to meet the challenges of the transition process - especially in the USA and the UK. Nevertheless, there is still a lack of research efforts and transition programs. In the light of the described risks in this patient group, efforts to improve transfer to adult healthcare are warranted.

## Engaging young adults in transitional and young adult kidney care services

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**BACKGROUND:** Patient survival rates for children with kidney disease are now currently 85-90% (Lewes et al 2008 and Ferris et al 2006) with many more young adults making the transition from paediatric to adult services. Poor transition from children's services may lead to disengagement with health services and can have serious outcomes for young people as well as incurring additional health service costs (Watson 2000). Engaging with young adults represents a particular challenge for health care professionals. Moving to adult units can be anxiety provoking, there may not be the same continuity of carers in adult services or accessibility to the same range of multidisciplinary professionals engaged in a young adults' kidney care. Isolation and loneliness may be exacerbated as young adults are invariably few in number. In addition these factors often combine with a time in a young adult's life where increased risk taking is prevalent and when young adults are yet to reach full cognitive maturity.

NHS Kidney Care commissioned and supported five project groups in England to develop new and innovative approaches to support young adults with kidney disease. In the London and South East project group, a survey was conducted at the lead London Trust, to identify the needs of young adults accessing adult kidney services.

**METHODS:** A survey, focusing on preparation for transition, clinic appointments, education and information, peer support and ways young adults keep in touch with their kidney services, were sent to 15-25 year olds with Chronic Kidney Disease 4 or 5, those with transplants or having dialysis. 54 surveys (a 61% rate) were returned.

**FINDINGS:** The survey has provided insights into young adults own recommendations for improving transition to adult services.

80% of young adults indicated they would attend a specific young adult clinic

50% of young adults had missed a clinic appointment

60% of those on dialysis and 40% of those transplanted would welcome the opportunity to speak with other young adults.

Only 7% of responses indicate that information provided in leaflets is helpful.

94 % own a mobile phone

70% would like to receive clinic reminders to their mobile phones.

CONCLUSIONS: In response a Multimodal Young Adult Clinics for 15-25 year old patients has been expanded, with increased multidisciplinary involvement. These clinics, importantly, encourage peer support amongst young people. Relationships between the paediatric and adult trusts are being strengthened in order to identify transitioning young adults earlier and ensure their attendance at young adult clinics. This has led to better communication and joined up working. All young adults receive a text reminder for this clinic and a parent area has been provided to facilitate communication and support for parents of transitioning young adults. To support such initiatives a young adult regional kidney care network, K2K, helps raise awareness of young adults in kidney services and provides online peer support for young adults by young adults. Young adults have also attended a national residential weekend, aimed at facilitating peer support and health promotion through activities and multidisciplinary educational workshops. The commitment continues to improve and expand the service in particular channels for patient education and information.

## “I feel like a freak – aren’t there any others like me?” The need for peer-support in paediatric nephrology

Jenny Prüfe, Centre for Family Research, Free School Lane, University of Cambridge, CB2 3RF, Cambridge, UK

**Background:** 999 children and adolescents in the UK received renal replacement therapy in 2010. Although this is a seemingly large number, these young people usually are the only ones in their home-environment to suffer chronic renal disease. Consequently, they have little contact with peers who find themselves in a similar situation.

**Aim:** To explore the needs for and chances of peer-interaction and peer-support in paediatric nephrology and discuss ideas based on suggestions from teenagers with chronic kidney disease stage 5.

**Methods:** A qualitative interview-study was conducted at a large teaching hospital in the UK. This part of the project involved 15 teenagers undergoing different forms of renal replacement therapies. Semi-structured interviews were transcribed verbatim and analysed using thematic analysis.

**Findings:** Participating teenagers expressed the desire to meet other young people with chronic kidney disease. This they argued would make them feel more similar to rather than different from their peers. Also, they hoped to prepare better for living with a chronic condition if they were able to learn about the experiences of other teenagers and see how they managed various disease-related situations. Teenagers undergoing dialysis particularly articulated the wish to meet with people who have already received a transplant and stressed how different it was to them to receive first-hand information as compared to the “theoretical knowledge” obtained from members of staff.

**Conclusions:** Teenagers wish to share their experience with peers who are in a similar situation as they are. However, these teenagers, especially those undergoing PD or pre-emptive transplantation, had little chance to meet each other. Suggestions which were made to change this included joint spare-time activities, group-holidays, self-help and discussion groups, as well as chat-rooms and online social-networking. The provision of peer-interaction and peer-support thus could add a valuable component to the psychosocial support of teenagers with chronic kidney disease.

## Adherence in paediatric renal failure and dialysis: competing responsibilities and ethical tensions

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Dr S Hulton (Consultant Paediatric Nephrologist, Birmingham Children's Hospital NHS Trust)

**Introduction:** Establishing and maintaining adherence in minors is problematic, especially in paediatric dialysis where the restrictions on fluids are extremely severe and breaches relatively easy for a minor to achieve. The severe regime is trying and its enforcement generates tensions for parents, minors and healthcare professionals (HCPs).

**Methods:** The aim of this study was to define and explore the ethical tensions for HCPs responding to nonadherence to the requirements for successful management of paediatric renal failure and dialysis.

11 nurses on a dialysis ward in a specialist children's hospital were interviewed about their experiences of nonadherence, responses to it, and views about the ethical tensions generated and how these could be resolved. Nurses were chosen as they work most closely with the minors and their families.

The interviews were audio-recorded, transcribed verbatim and analysed using conventional content analysis. The ethical tensions were also explored from a philosophical perspective so the results were informed by both theory and practice.

**Results:** Two overarching themes emerged: perceptions of nonadherence and responses to it. Participants felt that they, parents and minors (as they matured) all had responsibility for adherence, for different, overlapping reasons. They were sympathetic to the pressures that can lead to nonadherence but found it a difficult and frustrating problem to address. They took seriously their responsibility to protect the child's best interests and many thought this justified some coercion – e.g. enforced (against wishes of minor and/or parents) hospitalisation and overt disapproval (directed at both parents and more mature minors). They recognised that parents have competing obligations to other children, want to protect their own relationship with their child and that minors and their families long to be 'normal'.

Various responses to nonadherence were discussed: education, training, team approaches, coercion, hospitalisation, chastising, and persistence. The ethical dimensions and justifications were less well identified.

Discussion: The interests of the child are the primary concern for HCPs; but it is unclear how these are best promoted. A balance is needed between protecting a minor from immediate harm and equally detrimental damage to the HCP-minor relationship. Participants recognised that parents faced similar tensions (ensuring adherence whilst maintaining relationships) but not all were sympathetic and the behaviour of the minors was often described pejoratively. This may be due to concern to protect them from harm, as well as frustration. Scenarios were given that had clear ethical content that was not well articulated. Having the tensions clearly identified and defined may help HCPs cope with nonadherence, and provide tools for dissection, discussion and justification for the resolution of conflicts in the HCP-parent-minor triad. The usual mechanisms for dealing with refusal of medical treatment fit badly with what in other circumstances is both completely normal (e.g. drinking when thirsty) and legitimately governed by parental discretion. Despite its serious consequences, in this context, nonadherence may not concern medicines or treatment but the operation of the will and control of behaviour.

Conclusion: Explicit definition and exploration of the ethical tensions may help the HCPs to achieve a better understanding of their perceptions and responses.

## Managing short- term memory impairment in a child requiring renal transplantation: I know I'm angry but I can't remember why!

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**Introduction:** We describe the challenges of managing short- term memory impairment in the context of renal transplantation. Retention of information is crucial when preparing an adolescent for renal transplantation. A sixteen year- old girl known to our service experienced ongoing short- term memory loss following an acute neurological event. This created many unique challenges in supporting her psychosocial health during dialysis and around the time of transplantation. She could not recall important discussions and treatment plans as a result of her impairment. These included recalling names of hospital staff, benefits of transplantation, preparation for procedures and breaking bad news.

**Interventions:** We aimed to provide support through the period of preparation and eventual renal transplantation. We instituted a number of individual interventions within a systemic framework in order to address the challenges of short- term memory impairment. These included a photograph book of all staff, a personal copy of all printed material annotated by the patient, a diary to record the visits made by and the verbal information given by staff, recording all treatment decisions in clinical notes, and intensive input from a psychologist and play specialist.

**Conclusions:** Short- term memory impairment is an unusual challenge when preparing a child for renal transplantation. We have utilized a number of interventions in order to improve psychosocial support and facilitate patient empowerment.

## Chronic kidney disease (CKD) from the beginning of life: A challenge for infants, families and the multidisciplinary teams

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In this report a cohort of 21 children from the paediatric nephrology, the University Hospital of Cologne will be analyzed within their first three years of age.

All these patients as well as their families need a lot of support to cope with the early chronic kidney insufficiency, therefore the interdisciplinary care has been established in each paediatric nephrology in Germany. For this challenge, each interdisciplinary team is giving them the most professional kind of care. Even though, the outcome of the patients differs enormously. No one will be able to give an accurate prediction of the individual development of each patient, but the problems which occur are not only medical. Some families seem to need much more help, than we are able to give and at the same time they refuse the advice and support of the caregivers. Others just don't need that much help.

Quite often it seems to be much more difficult to cooperate with the parents than to take care of the medical problems of their children. More over: It might happen, that an interdisciplinary team will be forced to involve the youth welfare service against the will of parents, just to make sure, that a child is going to get the care that it needs. What does that mean?

What is to do, when all efforts of a modern health care system seem to overstrain the families?

To answer this question, it will need much more discussion and analysis of families and health caring systems within this course of disease. But with this small examination, some leading hints and signals might be found, that allow a better estimation of what kind of problems a multidisciplinary team for infants with CKD has to deal with.

Some milestones of early childhood development and dates of the mental development as well as medical and physical dates of this group of patients will be listed. On the other side, the subjective evaluation of the care givers applied to the communication, cooperation, adherence and outpatient support will be given.

## Life situation of adult patients with end-stage renal disease since childhood

Dominika Adamczuk, Maria Roszkowska – Blaim  
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Introduction: Advances in the treatment of end-stage renal disease (ESRD) in children resulted in prolongation of life, prevention of complications but also enabled returning to normal peer environment and active, satisfactory life.

Aim: Evaluation of life activity, disease acceptance (DA), satisfaction with life (SWL) and quality of life (QOL) in adult patients treated with RRT since childhood.

Patients and methods: 117 patients aged  $\geq 16$  years and treated with renal replacement therapy (RRT) since childhood were sent a set of questionnaires including three professional psychometric tools:

- a) disease acceptance (Acceptance of Illness Scale, AIS)
- b) life satisfaction (Satisfaction With Life Scale, SWLS)
- c) QOL (Kidney Disease Quality of Life, KDQOL)

and self-established survey (sociodemographic data, RRT modalities used).

The control group: 25 healthy subjects who were age- and gender-matched to the respondents in the study group.

Results: Return information was obtained from 71 (60.68%), including questionnaires from 45 respondents and information about death of 26 patients.

Patients who responded to questionnaires included 20 women and 25 men (age  $27.16 \pm 6.78$  yrs). Current RRT modalities included functioning kidney transplant (KTx) in 82.22% and hemodialysis (HD) in 17.78%. Mean duration of RRT was  $15.58 \pm 6.58$  years. KTx was performed in 97.78% of the respondents, including two KTx in 24.44%, and three KTx in 4.44%. During childhood, KTx was performed in 73.33%.

University education was reported by 18.18%, secondary by 50%, vocational by 13.64%, and elementary by 18.18%. Among the respondents, 46.67% were professionally active, 37.5% were still students, 62.5% were neither working nor studying. Social benefits due to inability

to work were reported by 66.67% of patients. 35% of women and 4% of men established their own families. Women were more likely ( $p<0.05$ ) to move out of the family home. Good disease acceptance was reported by 28.89% of respondents, poor by 26.67%, and average by 44.44%. The component of the AIS "I will never be self-sufficient in the way I would like to be" was rated the worst and correlated ( $r=0.47$ ) with the emotional component of the QOL. Worse life satisfaction ( $p=0.08$ ) was found in the study compared to the control group. No significant differences in regard to disease acceptance, life satisfaction, and physical and emotional components of the QOL were found between men and women, treated with HD vs KTx. Disease acceptance and the QOL were worse ( $p<0.05$ ) in patients living in rural vs. urban setting and in patients receiving social benefits. Age at the diagnosis of CKD correlated with life satisfaction ( $r=0.33$ ), time from the last change of the RRT modality with emotional well-being ( $r=0.34$ ). The number of KTx correlated negatively with the emotional component of the QOL ( $r=-0.66$ ) and emotional well-being ( $r=-0.73$ ).

#### Conclusions:

1. Education profile of adult patients receiving RRT since childhood is similar to the general population, but self-reliance in life is low, as evidenced by rare establishment of own families, often unemployment and frequent living together with parents.
2. Good disease acceptance was reported by 1/3 of patients receiving RRT since childhood, with a lack of self-sufficiency being a major problem. Emotional well-being was negatively affected by young age at the time of CKD diagnosis, experience of Tx loss, receiving social benefits, and rural background.

## “As a parent you have no choice” – decision making processes in the context of living related kidney donation

Jenny Prüfe, Centre for Family Research, Free School Lane, University of Cambridge, CB2 3RF, Cambridge, UK

**Background:** Each year numerous parents in Europe face the question whether or not to donate one of their kidneys to their child. Regulations for living donations are strict and framed by complex ethical guidelines. Above all, the potential donor's decision needs to be a fully informed one, ensuring that all risks and benefits are understood. Consent must be voluntary and free of coercion. However, if a parent's own child is in dire need of an organ, can we really consider this to be a decision free from any duress?

**Aim:** To explore parents' decision-making processes in the context of living related kidney donation and analyse its consequences for clinical practice.

**Methods:** A qualitative interview-study was conducted at a large teaching hospital in the UK. This part of the project involved nine mothers and five fathers whose child was either on-call, scheduled for living-donation, or had received a transplant no more than 12 months prior to data-collection. Semi-structured interviews were transcribed verbatim and analysed using thematic analysis.

**Findings:** Parents expressed two types of decision making: either they decided instantaneously based on an emotional response and often before the topic was raised by members of staff, or they announced their decision after careful consideration of pros and cons. However, it was frequently stressed that the rational process became a rather emotional one if parents received the information that - from a medical perspective - living-donation is likely to provide the better outcome: e.g., “Once you are told ‘this is the best for your child’, there is not really a choice anymore – as a parent you always want to do the best.”

**Conclusions:** The nature of the parent-child relationship in itself might significantly influence parental decision-making. While parents are unlikely to experience any duress from staff or family-members, they feel a moral obligation as a parent to do what is best for their child.

We need to acknowledge this internal pressure to provide adequate support for the families concerned.

## Reintegrating Children and Adolescents with Nephrotic Syndrome

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**Introduction:** "Arabkir" JMC&ICAH is the Republican reference centre for children and adolescents with various renal diseases. The psychosocial and hospital school services have been introduced and developed here since 1991 as part of the holistic approach in paediatrics. Today, the hospital school is run and psychosocial services are provided to the patients in cooperation with DAA, local NGO.

**Situation:** Doctors pay close attention to the nephrotic syndrome in children and adolescents. The number of patients with progressing illness doesn't decrease and often it becomes resistant to therapy. Such patients receive special disability status. The clinicians pay attention mainly to the hospitalization stage. Studying the quality of life of these patients isn't a less important issue of modern pediatrics. Though there are no criteria and standards to estimate the quality of life, the systematic multidisciplinary approach towards increasing the efficiency of the treatment and solving special social problems related to the illness is essential.

**Problem:** Due to the scarcity of resources in Armenia patients from the regions are hospitalized for longer periods and need also psychosocial support. DAA runs hospital school where school age children catch up with the school program while on treatment. The educational developmental games for all age children are used both for teaching and entertaining. On the grounds of the hospital there is a handicraft atelier with different workshops that allow the patients to fill their hospital routine with interest and feel socially adapted.

At present one of the problems of the patients with nephrotic syndrome is that their home school teachers are uninformed of their condition.

**Solution/Conclusion:** One of the most important points is to inform the home school teachers about the illness and its consequences in order they can organize the reintegration of these patients. This can be done through:

- special workshops in cooperation with hospital school teachers, e.g. Special Educators' Club
- visits of the hospital school teachers to home schools and vice versa.

## Psychosocial aspects of children with disorders of sexual development (DSD)

L. Marshall, Lübeck

The care for patients with DSD and their families demands a wide spectrum of psycho medical offers depending on DSD type, time of disclosure and family resources. In the past, diagnosis and medical treatment was handled solely without considering the patients will or providing him with sufficient information. For many years self help groups have complained about and fought against this approach.

Today a multidisciplinary team (MDT) strives for an Informed Consent which encompasses understandable disclosure and outlining the array of treatment possibilities. Irreversible decisions such as surgical interventions should be postponed to await the patients consent. A MDT should be defined in tertiary carecentres. The team should offer the expertise of a paediatric specialist in endocrinology, surgery or urology, psychologist and genetics.

The key elements of psychomedical management in DSD are the following:

1. Support of the family with the newborn in order to promote positive adaption
1. Supporting the disclosure process with open communication
2. Psychological assessment of gender identity in the patient
3. Long term care according to the patients and families needs.
4. Facilitate team decisions about gender assignment, diagnostic procedures, timing of surgery, and sex hormone replacement therapy
5. Enhancing quality of life encompassing falling in love, ability to develop intimate relationships, sexual functioning, the opportunity to marry and to raise children, regardless of biological indicators of sex.

### Gender assignment in newborn infants

Initial gender uncertainty is very stressful for the family and demands a good support from the very beginning. The team needs to provide a diagnosis as soon as possible to reduce agonizing waiting time and uncertainty. Except for the life threatening, salt loosing form of congenital adrenal hyperplasia the situation is mostly not a medical emergency and should be dealt with thoughtfulness and calmness. Parents need to be equipped with a roadmap that highlights times when parenthood should focus on the strength and normality of their child and times in which sensible windows charge reconciliation and a step in the decision process.

The psychosocial team offers dialogue to bridge the time of waiting, to express fears about the consequences and to accompany the family over a long period of life. It should help building a net for the family, that may arrange psychosocial support close to home and connect to support groups and arrange contacts to other DSD parents or patients.

### Disclosure in the adolescent

Teenagers contact us under various conditions. Often the absence of menorrhoea leads to the referral to the DSD Centre. Comparison in the physical development to their peers and information through the internet sometimes have led to disturbing hypothesis. The disclosure of a diagnosis snowballs a number of issues and concerns on both patients and parents side. Questions about identity and sexual identity, partnership, fertility, risk for malignancy suddenly bring the perspective of an unburdened life to an abrupt end.

Under this condition achieving Informed Consent means supporting the mature patient and his parents to discuss and weight all options. These might span from promotion of a positive adaption, Timing of further diagnostic investigation, surgery, sex hormone replacement therapy and mediating local psychotherapy. Patients also use our service to communicate their situation towards their partners.

The DSD Centre in Lübeck also serves the demand for clinical studies from the German ethic commission in Feb 2012. Prospective studies, as well as research on resilience factors for therapeutic support need to enhance the patient centered care in DSD.

## Genital malformations and disorders of sex development – a subanalysis of the German WT1-Registry

Anja Lehnhardt, Claartje Karnatz, Gabriele Holst, Markus Kemper

Introduction: Chromosomes, hormonal profiles, internal and external sex organs define male and female sex. Disorders of sex development (DSD) can be caused by chromosomal and hormonal abnormalities and result in genital ambiguity and incongruent combinations of gender-specific external genitalia, reproductive ducts and gonads. DSD should not to be confused with 'gender dysphoria', which is also known as transsexuality or being transgender. Gender dysphoria is where the reproductive organs and genitals have developed normally, but the person feels that their gender identity (their sense of what sex they are) does not match their biological sex.

The Wilms Tumor Suppressor Gene 1 WT1 encodes for a transcription factor, which plays an important role in regulating the development of the urogenital system. WT1-Germline mutations classically lead to two different syndromic diseases, Denys-Drash Syndrome (DDS) caused by exon-mutations and Frasier Syndrome (FS) caused by intron mutations.

Method: Our registry is an initiative of the German pediatric renal society (GPN). We aimed to retrospectively analyse all patients with a known WT1-germline mutation from German-speaking countries. Obviously the main focus is on renal phenotype, development of proteinuria and endstage renal disease. We additionally gained detailed data on genital malformations, sex and gender as both syndromes include 'male pseudohermaphroditism' in their classical definition, today we use the phrase 'DSD with 46,XY karyotype' („46,XY DSD“ )instead.

Results: So far 42 patients from 16 centres were included. In 34 patients an exon mutation was found. 35 did have proteinuria, 16 patients had Wilms Tumors, 30 patients required renal replacement therapy. 24/42 patients showed genital malformations (main diagnoses: severe hypospadias, kryptorchidism, mikropenis) requiring n=61 operations. 19/42 patients have 46, XY DSD, 3/42 patients have complete gonadal dysgenesis (all 3 patients with intron-mutations). 25/42 had XY karyotype, 15/42 XX karyotype, 6 XY-patients are raised with a female gender.

Conclusion: Patients with WT1-germline mutation do not necessarily have genital malformations or DSD. If so, they do tend to have 46, XY DSD with combined malformations (Exon-Mutations) or complete gonadal dysgenesis (Intron-Mutations) requiring repeat surgery. It is not always easy to delineate DSD from 'simple' genital malformations, and paediatricians should be aware that DSD does not exclusively describe the new born with ambiguous genitalia but can also become obvious later in childhood and puberty (e.g. in hypogonadism after maldescensus testis). Pediatric patients with DSD should be checked for proteinuria and if positive, WT1-mutations should be considered. Phrases such as 'Intersex' or 'Hermaphroditism' must not be use anymore.

## Assessment of Health-related Quality of Life in Children with End-Stage Renal Disease

U. Ravens-Sieberer, Hamburg

The health-related quality of life (HrQOL) of children and adolescents is increasingly considered a relevant topic for clinical research. Instruments to assess quality of life in children and adolescents of a generic as well as disease- or condition-specific nature are being developed and applied in epidemiological surveys, clinical studies, quality assurance and health economics. This presentation attempts to give an overview on the state of the art of HrQOL assessment in children with chronic conditions such as end stage renal disease as it relates to methodological and conceptual challenges. Instruments available in international or cross-cultural research to assess HrQOL in generic terms are identified and described according to psychometric data provided and the width of application. Several challenges in the assessment of child and adolescent HrQOL are identified, ranging from conceptual and methodological to practical aspects. Five specific major issues are considered: (i) What are the dimensions of HrQOL relevant for children and adolescents with chronic conditions, and do suitable instruments for their measurement exist? (ii) Can these dimensions be collected in a cross-culturally comparable way? (iii) What advantages and disadvantages do self-rated versus externally evaluated HrQOL measurements of children and adolescents have? (iv) How can HrQOL be assessed in an age-appropriate way? (v) What are the advantages and disadvantages of disease-specific and generic data collection? Future research in measures of quality of life in pediatric patients will be introduced to assist health professionals in making more informed clinical decisions, using patient-centered assessments.