MEDICAL UNIVERSITY OF VIENNA

Be on TRAQ – Cross-cultural adaptation and pilot-testing of the Transition Readiness Assessment Questionnaire for use in German speaking Youth with Special Health Care Needs

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Submitted by

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Declaration

Herewith I declare that I have written original work independently and that I am the sole author of this thesis. The thesis was conducted under the supervision of Ao. Prof.in Gabriele Häusler at the Department for Pediatrics and Adolescent Medicine of the Medical University of Vienna. The information reported is the result of my own work and studies, except where due to reference is made. I have followed the rules of good scientific practice to the best of my knowledge.

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Abstract in English

Background: Internationally, the change from comprehensive pediatric care to specialized adult health care, the so-called health care transition (HCT) is associated with detrimental effects on health outcome in young adults with chronic health conditions. HCT poses a variety of challenges for adolescent patients with a chronic condition, their parents, health experts and the medical system per se. For young patients diagnosed with a rare disease, HCT is particularly challenging. Standardized processes are often lacking. The release from pediatric care is usually scheduled around the age of eighteen. Instead of focusing on birthdates, a growing body of literature recommends the assessment of readiness for transition.

Aims: The main aim of this thesis was to develop a generic transition readiness assessment questionnaire suitable for German speaking youth with special health care needs (YSHCN).

Methods: After comprehensive literature research on health care transition and a thorough investigation of transition services, in particular for patients with a rare disease, we translated and cross-culturally adapted the American assessment questionnaire, the TRAQ 5.0. The pilot-testing of the instrument comprised the administration of the German version to YSHCN. Psychometric properties were statistically analyzed. Feasibility was tested regarding to how much time was needed or how often help was provided in the course of filling out the questionnaire. In a next step possible differences in transition readiness scores were investigated between a patient group diagnosed with Turner syndrome (TS) and a group of female patients with type 1 diabetes or with a rheumatic condition, respectively.

Results: This work resulted in the German version TRAQ-GV-15, comprising 15 items and 3 subscales “autonomy”, “health literacy”, “adherence”). The TRAQ-GV-15 presented acceptable psychometric qualities, comparable to other cross-cultural adaptations of the TRAQ 5.0. Internal consistency with an overall Cronbach’s alpha of 0.824 indicates a good reliability. Age, in contrast to sex, had a significant effect on the TRAQ scores. In addition, there are indicators that the diagnosis of TS might be responsible for lower TRAQ-GV-15 scores compared to type 1 Diabetes or rheumatic

diseases. In addition, patients with TS needed significantly more time to fill out the TRAQ-GV-15.

Conclusions: The TRAQ-GV-15 is the first generic transition assessment instrument for German speaking YSHCN with reported psychometric properties as far as we know. The administration of the instrument is time-efficient and feasible in resource-limited clinical settings. The TRAQ-GV-15 could help to identify potentials for patient education and to facilitate the evaluation of transition services and of follow-up research in the field of HCT.

Abstract in German

Hintergrund: Der Übergang von pädiatrischen Zentren, Kinderkliniken, in die medizinische Betreuung für Erwachsene, die sogenannte Transition, ist international für die meisten jugendlichen Patientinnen und Patienten mit chronischer Erkrankung eine große Herausforderung. Leider zeigte sich in den letzten Jahrzehnten, dass sich die Gesundheit junger Erwachsener nach der Transitionsphase massiv verschlechtert. In dieser insgesamt fordernden Lebensphase stellen sich auch für ihre Eltern und das betreuende ExpertInnenteam viele Fragen. Für junge PatientInnen mit einer seltenen Erkrankung können die Hürden in der Transitionsphase sogar noch größer sein. Bis jetzt gibt es wenig standardisierte Prozesse, Abläufe und Zuständigkeiten sind teilweise unklar. Dennoch werden PatientInnen üblicherweise rund um den 18. Geburtstag von den pädiatrischen Zentren verabschiedet. Im Zuge der Auseinandersetzung mit Verbesserungen im Transitionsprozess wurde in den letzten Jahren verstärkt die Feststellung der Bereitschaft von Jugendlichen zur Transition mit geeigneten Fragebogen empfohlen.

Ziel: Das Hauptziel dieser Arbeit war die Übersetzung und kulturelle Adaptierung eines international angesehenen und krankheitsneutralen Fragebogens, der die Bereitschaft von chronisch kranken Jugendlichen zum Wechsel in die Erwachsenenmedizin erfasst. Weiters sollte überprüft werden, ob es Hinweise auf Unterschiede in Bezug auf Transitionsbereitschaft zwischen Diagnosegruppen gibt.

Methodik: Im Zuge einer umfassenden Literaturrecherche zum Thema Transition, der internationalen Versorgungslage im Bereich Transition, Transition in Bezug auf seltene Erkrankungen und zu Transitionsprogrammen, stießen wir auf den gut untersuchten und validierten amerikanischen Fragebogen TRAQ 5.0. Nach Übersetzung und Adaptierung für den deutschsprachigen Raum wurde die deutsche Version an 172 jugendlichen PatientInnen mit unterschiedlichen chronischen Erkrankungen pilotiert. Die statistischen Verfahren umfassten Faktorenanalyse, Regressionsanalysen und deskriptive Statistik. Interne Reliabilität und Validität wurden überprüft und beschrieben. Zusätzliche wurden die benötigte Durchführungsdauer und Beratungszeit sowie die Anzahl an Hilfestellungen zur Beantwortung des Fragebogens dokumentiert. In einem nächsten Schritt wurden 27 Patientinnen mit Turner syndrome (TS) mit 27

Patientinnen mit entweder Typ 1 Diabetes mellitus oder juvenilem Rheuma in Bezug auf TRAQ-GV-15 Werte verglichen.

Ergebnisse: Aus dieser Arbeit resultiert die deutsche Version TRAQ-GV-15 mit 15 Fragen (Items), die sich in drei Subskalen („Autonomie“, „Gesundheitskompetenz“, „Adherenz“) zusammenfassen lassen. Der TRAQ-GV-15 zeigt akzeptable psychometrische Qualitäten, die sich mit den anderen international adaptierten TRAQ 5.0 Versionen auf spanisch, brasilianisch und türkisch vergleichbar sind. Die interne Konsistenz mit einem gesamten Cronbach’s alpha von 0.824 deutet auf eine gute Reliabilität hin. Im Gegensatz zum Geschlecht hatte das Alter einen signifikanten Einfluss auf die TRAQ-Ergebnisse. Zusätzlich gab es Hinweise, dass die Diagnose von TS für geringere TRAQ Ergebnisse verantwortlich sein könnte, verglichen mit den TRAQ-Werten von Teilnehmerinnen mit den Diagnosen Diabetes Typ 1 oder einer juvenilen rheumatischen Erkrankung. Teilnehmerinnen mit TS benötigten signifikant mehr Zeit zum Ausfüllen des Fragebogens und erreichten signifikant niedrigere Werte im TRAQ.

Schlussfolgerungen: Der TRAQ-GV-15 ist der erste krankheitsneutrale Fragebogen zur Erfassung von Transitionsbereitschaft chronisch kranker Jugendlicher in deutscher Sprache soweit wir wissen. Der Einsatz des Instruments ist zeitsparend und leicht durchführbar, auch in ressourcenbewussten klinischen Zusammenhängen. Der TRAQ-GV-15 könnte sowohl die Feststellung von Schulungsbedarf, sowie die Evaluierung von Transitionsprozessen und –angeboten sowie Langzeitstudien in der Transitionsmedizin begleiten und erleichtern. Ich hoffe, mit dieser Arbeit einen Beitrag zur verbesserten Versorgung von jugendlichen PatientInnen in der Transition zu leisten.

Publications arising from this thesis

Care of girls and women with Turner syndrome: beyond growth and hormones.

Culen C, Ertl DA, Schubert K, Bartha-Doering L, Haeusler G.

Endocr Connect. 2017 May;6(4):R39-R51. doi: 10.1530/EC-17-0036. Epub 2017 Mar 23. Review.

PMID: 28336768

Be on TRAQ — Cross-cultural adaptation of the Transition Readiness Assessment Questionnaire (TRAQ 5.0) and pilot-testing of the German Version (TRAQ-GV-15)

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Less ready for adulthood? – Turner syndrome has an impact on transition readiness

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Manuscript in preparation

Additional publication in the context of this thesis:

Health status, quality of life and medical care in adult women with Turner syndrome.

Ertl DA, Gleiss A, Schubert K, **Culen C**, Hauck P, Ott J, Gessl A, Haeusler G.

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Abbreviations

AYA Adolescents and Young Adults
ASD Autism Spectrum Disorder
BTM Berliner Transitionsmodell
DKA Diabetic Ketoacidosis
DRK Deutsches Rotes Kreuz
EFA Exploratory Factor Analysis
GHD Growth Hormone Deficits
GV German Version
HCP Health Care Providers
HCT Health Care Transition
HRQoL Health Related Quality of Life
IBD Inflammatory Bowel Disease
JIA juvenile Idiopathic Arthritis
QoL Quality of Life
SCD Sickle Cell Disease
SMS Short Message Service
T1DM type 1 Diabetes mellitus
TRAQ Transition Readiness Assessment Questionnaire
TS Turner Syndrome
UK United Kingdom
YSHCN Youth with Special Health Care Needs

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CHAPTER ONE: INTRODUCTION

1.1. General introduction

About five years ago, our study group decided to invite former patients treated at the Pediatric Outpatient Clinic for Endocrinology for long-term follow-up. Main research objectives were current health status and Quality of Life (QoL) in adult women with Turner syndrome (TS) (Ertl et al., 2018). In the course of the study it became apparent that the women were experiencing a poor standard of medical care. This realization has led to an intensive research process in the field of transition from pediatric care to the adult health system in general and specifically in the field of rare diseases. We started exploring TS specific challenges and impediments and searched the literature for recommendations on well-prepared transition processes. Transition related efforts started earlier in the Anglo-American health systems than in European countries. However, international endeavors have followed in the past years, contributing to a growing body of scientific literature. In addition to standardized transition programs, questionnaires are available, developed for patients, caregivers and health experts, either for baseline assessment within the transition process or aiming at enhanced patient activation. These days, the implementation of standardized transitional processes is under way worldwide. Nevertheless, the situation in Austria could be improved.

Although there is a general sense that improvement of the transitional care is of great importance, defined care pathways to minimize the risk for getting lost-for-follow-up after being released from pediatric care are still lacking. Pilot-projects have been carried out, however, no best practice model has been established at our clinic, or in Austria respectively. In our search for guidelines for enhanced transfer processes we came across the term transition readiness and transition readiness assessment tools, which caught our interest. The outlook of preparing patients with rare diseases moving to adult care from early age on was promising. Then, no generic transition questionnaire or validated assessment tool for use in the transitional phase in German

language was available. With a population of approximately 100 million people, of which twenty per cent are youth under the age of 18, an estimated fifteen to twenty per cent of the adolescents are diagnosed with a chronic disease (“Eurostat,” 2019). This is a relevant, not negligible patient group.

We searched for internationally renowned questionnaires suitable for cross-cultural adaptation. It was not long before we encountered a well-studied and disease-neutral transition readiness assessment tool, the Transition Readiness Assessment Questionnaire (TRAQ) 5.0 (Wood et al., 2014). The TRAQ 5.0 was developed by pediatricians in the United States (US). The questionnaire comprises 20 items in five subscales. Compared to other transition readiness assessment instruments the TRAQ 5.0 was the best-validated tool (Zhang, Ho, & Kennedy, 2014). It has sound psychometric qualities such as high reliability (overall Cronbach’s alpha .94) and good validity.

In my thesis, first, I would like to present an overview on the current knowledge in the field of transition research. Second, I will introduce the background for my studies to translate, cross-culturally adapt and pilot-test the TRAQ 5.0 for use in German speaking countries. This step included a comparative study between patient groups diagnosed with TS, type 1 diabetes and a childhood-onset rheumatic condition. Third, I will present my studies in the result section. Last, I will discuss the results of the conducted work and give an outlook on potential future developments.

1.2. Health Care Transition (HCT)

Due to advances in medical research and treatment the majority of children and adolescents diagnosed with a former life-threatening now chronic condition survive into adulthood (*Cystic Fibrosis Trust*, 2008; Watson et al., 2011; Bryant et al., 2014; Campbell et al., 2016). In Austria, estimated 190.000 children and adolescents suffer from a chronic physical condition (Fichtenbauer, 2015). With older age prevalence of chronic diseases increases, ranging from 13% in ten- to eleven-year-old children up to 20% in seventeen-year-olds (Hölling et al., 2012). Additionally, special attention should be paid to the cohort of children and adolescents with rare diseases, estimated 4.000 in Austria ("Www.eurordis.org," 2019; "Www.prorare-austria.org," 2019). Assessment indicates that not all chronically ill adolescents are in the need of elevated levels of care or nursing in adulthood (Klimont & Baldaszti, 2014). Nevertheless, youth with special health care needs (YSHCN) have to change their health care providers. Transitioning to adult medical health care systems for long-term medical surveillance applies to most of them. Yet, they face a bundle of difficulties along their way. Transition proved to be challenging for all chronic conditions.

Blum´s definition from the year 1993 still applies in the opinion of most researchers and clinicians. Blum defines transition as "the purposeful, planned movement of adolescent and young adults with chronic physical and medical conditions from child-centered to adult-orientated health care systems" (Blum et al., 1993). Whereas the term "transfer" simply refers to the change of facility or location where medical care is provided, e.g. from comprehensive pediatric clinics to adult medical care (Fredericks, 2017).

The success story in pediatric care has led to vast improvement in higher survival rates as well as in decreased morbidity and mortality in chronically ill children and adolescents (Rosen, Blum, Britto, Sawyer, & Siegel, 2003; Viner, 2008). Children born today with cystic fibrosis have good chances to survive into their fifties (Dodge, Lewis, Stanton, & Wilsher, 2007; Tuchman, Schwartz, Sawicki, & Britto, 2010), meaning the proportion of individuals with cystic fibrosis achieving adulthood (older than 18 years of age) increased from 27% to 56% (*Cystic Fibrosis Trust*, 2008). More than ninety percent (93%) of the patients diagnosed with sickle cell disease (SCD) will also survive childhood and adolescence (Bryant et al., 2014). In Nephrology, literature

reports survival rates of 85–90% (Lewis & Slobodov, 2015). Almost 90% of children with congenital heart disease will survive into adulthood (Moons, Bovijn, Budts, Belmans, & Gewillig, 2010).

Nevertheless, clinicians realized poor health outcomes in YSHCN after leaving comprehensive pediatric care (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011). Transition has been identified as a risk period for disengagement, loss to follow-up, increased health risks and decreased QoL in all kind of childhood onset chronic illnesses. As an example, in Germany only one third of adult patients with CF are followed up in an adult center (Fischer, Nährig, Kappler, & Griese, 2009).

High risk for detrimental health during transition is known for type 1 diabetes (Cameron, Amin, de Beaufort, Codner, & Acerini, 2014; Delamater, 2007), cystic fibrosis (CF) (Fischer et al., 2009), juvenile idiopathic arthritis (JIA) (White & Ardoin, 2015), inflammatory bowel disease (IBD) (Keller, 2010), congenital heart diseases (CHD) (Sable et al., 2011) and many more. For rare diseases such as haemophilia (Breakey, Blanchette, & Bolton-Maggs, 2010), for endocrine syndromes such as TS (Gleeson, McCartney, & Lidstone, 2012; Gawlik et al., 2012; Ertl et al., 2018) or Prader-Willi syndrome (Paepegaey et al., 2018), for congenital adrenal hyperplasia (ADG) (Bachelot et al., 2017) and for diseases including growth hormone deficiency (GHD) (Courtillot et al., 2013) worrying health outcomes have been reported after being transferred. Medical parameters decline, therapy regimen adherence decreases, non-compliance regarding medication intake increases. For instance, one year after transfer of adolescents and young adults (AYA) with diabetes Hba1c increased significantly, incidences of Diabetic Ketoacidosis (DKA) with hospitalization and severe hypoglycemia doubled (Walleghem, MacDonald, & Dean, 2006). Microvascular complications in diabetes are of detrimental effect on health outcome (Kappelen et al., 2018). An increased number of loss of transplanted organs are observed in young adults after transplants (Watson et al., 2011).

Transfer from highly supportive, child and family centered pediatric care to adult individuals centered health care is theoretically scheduled by the age of eighteen (American Academy of Pediatrics, American Academy of Family Physicians, and

American College of Physicians, Transitions Clinical Report Authoring Group, 2011; Betz, 2004; Kreuzer et al., 2015; Sheehan, While, & Coyne, 2015). However, actual transfer of chronically ill AYA sometimes is delayed (Lotstein, 2013). It comes along with a vulnerable time in the lives of adolescents. Manifold challenges are posed on AYA, involving physical, cognitive and emotional development that affect about every aspect of their lives. In addition, maturation in these different developmental areas mostly do not happen simultaneously (Sanders, 2013). Furthermore, only about 18% of the youth and 27% of the parents reported plans on HCT (Sawicki, Kelemen, & Weitzman, 2014). Therefore, within the last decade, the topic of HCT has become a major issue in health care services not only in the Anglo-American countries, but also in Europe. Initiatives lobbied for enhanced transition for chronically ill adolescents as well as for handicapped adolescents in German speaking countries ("Transition1525," 2019; "Zone," 2019). Research increasingly placed the focus on transition processes within the last years (Pape & Oldhafer, 2017). Nevertheless, research pinpoints that upcoming transfer often had not been addressed in conversations with YSHCN (Jensen et al., 2017; Stewart et al., 2017). Additionally, recent data from the USA disclose that still 85% of AYA do not receive transition services from their medical care providers (Lebrun-Harris et al., 2018).

In Austria, the need for improved transition was recognized years ago (Deutsch, Gobara, & Waldhauser, 2016). The example of long-term care of survivors of childhood-onset oncological diseases quite clearly show the lack of systematic follow-ups and missing clinicians in charge. A comprehensive model of care, derived from a multi-professional Delphi-Process, was presented not long ago. Recommendations comprise the development of a registry, case coordination and case management. Setting up of a nationwide network of experts should support structured longitudinal studies as well as quality control ("Zone," 2019).

Notwithstanding this fact, transition activities at present still depend on committed pediatricians and collaborative colleagues in the adult setting. No standardized overarching transition policy has been implemented to date. Transition services are not mandatory. Further, reimbursement of transitional services is nonexistent so far.

1.3. Research on HCT service

In 2004, researchers in Los Angeles stated a growing need for transition service evidence. Their review of 43 studies published over a period of 20 years indicated that transition intervention research was still in its early stages. By then, outcome research in the field of transition was characterized by a lack of theoretical framework, by inadequate research designs and by the absence of valid and reliable tools (Betz, 2004). Some years later, paucity in evidence for best practice, about both process and uncertainty which factors determine effective care during the transitional phase remain (Watson et al., 2011, 2011) (Watson 2011). Data on transitional processes, best practice and transition outcome are still scarce (Ludvigsson et al., 2016). However, there are indicators that the situation is improving or at least that major efforts for improvement are under way.

There is evidence that since 2010 health outcomes after structured and multidisciplinary intervention programs have improved, at least for young adults with CF or type 1 diabetes. In an updated comprehensive review, researchers analyzed a final of 61 studies which indicated improvement in transition outcomes especially for patients with CF and type 1 diabetes (Zhou, Roberts, Dhaliwal, & Della, 2016). Another review evaluated the effectiveness of interventions that aim to improve the transition process (Campbell et al., 2016). Due to the limited number of reported interventions, only four studies met the inclusion criteria. Two studies showed small positive effects on transition outcomes. A randomized controlled trial (RCT) with 81 participants aged twelve to twenty evaluated a technical approach to transitional care over an 8-month period. YSHCN in the intervention group received disease related management advice via tailored SMS. Positive effects were reported for two and eight months follow-up (Huang et al., 2014). Another study investigated the effects of a one hour nurse-led intervention with adolescent patients aged 15 to 17 addressing cardiac knowledge and disease self-management (Mackie et al., 2014). Overall effects of the evaluated studies showed small effects in enhanced transition outcomes such as increased disease knowledge, improved quality of life or enhanced health status (Campbell et al., 2016).

Improvements regarding more regular clinical attendance, improved satisfaction with care and decreased diabetes-related distress in AYA with type 1 diabetes were

reported from a transition program that included a transition coordinator (Spaic et al., 2019). In line with these results, the implementation of a comprehensive, structured transition program for CF patients in San Francisco led to improved outcome after an 18-months intervention (Okumura et al., 2014). Opposed to these findings, a Canadian group reviewed transition programs for patients with epilepsy from around the world. Research objective was to examine the models of transitional care for AYA with epilepsy in five countries (Canada, Colombia, France, German und the United Kingdom). The five models of care show considerable differences. The investigated services ranged from joint clinics in the UK and Canada to implementation of checklist in the pediatric setting. Comprehensive evaluation is still lacking. No decision on which of the five transition initiatives could serve as best-practice model could be made. No evidence for effectiveness was found to date due to lack of well-designed evaluation studies (Camfield et al., 2019; Carrizosa, An, Appleton, Camfield, & Von Moers, 2014).

To date, most intervention studies have been investigating on a patient level, using educational programs, workshops or questionnaires. Interventions on a systemic level, addressing the organizational side of transition, were neglected (Campbell et al., 2016). An Australian group described the need to also focus on health related quality of life (HRQoL) and overall QoL, instead of solely having the eye on health outcomes (Sawyer & Ambresin, 2014). Individual and social outcome instead of health services outcome are required elsewhere (Fair et al., 2015). Instruments such as the “Mind the gap scale” (Shaw, Southwood, McDonagh, & the British Society of Paediatric and Adolescent Rheumatology, 2007) aim at surveying patients’ and caregivers’ satisfaction with transitional services. However, challenges in conducting well-designed studies in the field of transition are related to scarce resources regarding personnel, time and money.

Despite the endeavors seen in the last years, one can state, transition intervention research is still in its infancy (Gabriel, McManus, Rogers, & White, 2017). Joint clinics to facilitate cooperation appear to be cost-intensive (Carrizosa et al., 2014), however, studies on cost-effectiveness are scarce and transition programs in general are not well evaluated (Nabbout et al., 2019). Furthermore, the quality of methodology for longitudinal studies is poor (White & Cooley, 2018) and there is still no evidence for the sustainability of positive effects (Spaic et al., 2019).

The triple aim framework helps to evaluate public health actions regarding the three aims population health, patient experiences and costs. Prior and colleagues (Prior, McManus, White, & Davidson, 2014) criticized that few studies comprehensively evaluate transition in relation to efficacy as measured by the triple aim concept. Interventions attempt to improve transition, but a set of adequate measures is needed to evaluate and compare intervention outcomes.

A review with regard to the triple aim framework investigated transition in primary health care settings. Only three studies met the inclusion criteria. Thus, the authors concluded that very limited empiric evidence is available. In contrary to transition studies in clinical settings, cost domain was the most commonly reported outcome, but no empiric evidence was provided for improved transition in the primary care sector (Bhawra, Toulany, Cohen, Moore Hepburn, & Guttmann, 2016).

A more fruitful review revised forty-three transition related studies also within the triple aim framework. Statistically positive outcomes were found in twenty-eight studies. Twenty studies reported positive outcomes related to population health. Eight studies described positive patient experiences. Evaluation of costs were evaluated in only three studies. However, no significant savings were found. Further, adherence to therapy regimen or adherence to care as well as utilization of ambulatory care in adult settings were commonly reported as outcome parameters (Gabriel et al., 2017).

1.3.1. Parameters for successful HCT

The search for parameters for successful transition reveals challenges and barriers that are common for all chronic conditions as well as disease specific challenges. Researchers conducted a Delphi-study to identify parameters relevant for HCT outcome. Within a three-step-process, YSHCN, parents and caregivers as well as health care providers ranked a large number of items, associated with the HCT process. Parameters rated as relevant for successful transitioning included individual outcome (e.g. QoL, disease knowledge and self-management skills), social outcome (e.g. having a social network) as well as health related outcomes (new medical home). In this study, however, long-term health outcome has been neglected (Fair et al., 2015).

The observation above is underpinned by another study. Transition research in patients with type 1 DM often focuses on transitional processes and neglects transition outcomes. From qualitative interviews with patients, experts, and caregivers the authors derived measurements on transition outcomes such as biomedical markers, disease specific knowledge and management skills. Further, navigation of a new health care system, balance of parental involvement and autonomy were announced as key parameters for successful transition (Pierce et al., 2017). These findings go in line with a prospective evaluation which found good transition outcome associated with only three elements: appropriate parental involvement, meeting the adult team before transfer and promoting health self-efficacy (On behalf of the Transition Collaborative Group et al., 2018).

1.3.2. Barriers for successful HCT

The authors of a comprehensive review described the following five barriers prior to transition or in the course of transitioning: One, patients experience inadequate preparation before their transition. Second, access to adult care providers represents a big challenge. Third, complex health condition, eventually comprising cognitive impairment, hamper the transitional process. Four, parental attitude toward the transfer impedes a smooth shift. Last, the reluctance of pediatric providers to hand over their patients might also attribute to difficulties (Zhou et al., 2016).

Another recent review on barriers for successful transition confirms the above identified problems. Limited access to specialized adult care as well as insufficient disease related knowledge and lack of skills regarding self-organization were reported. Negative beliefs and poor expectations were also associated with the upcoming transition phase. The most challenging process was seen in the change of health care providers. The letting-go of long-lasting and trustful relationships between pediatric patients, their families and the health care team seems to be a universal difficulty (Gray, 2018). This goes in line with a very recent study from New Zealand, where the main concern of AYA with IBD articulated in focus groups was the building of a new relationship with a new health care provider team (Karim, Porter, McCombie, Gearry, & Day, 2019).

Hence, barriers to transitioning can be noted on the level of patients, of caregivers and of providers. Caregivers and pediatric health care providers (HCP) often are insecure about when is the right moment to shift responsibilities from caregivers to AYA (Fredericks, 2017). On the level of adult providers both a lack of availability and a lack of training in congenital and childhood-onset conditions for adult providers can be stated (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011; Lemly, Weitzman, & O'Hare, 2013; Bryant et al., 2014). Furthermore, difficulties in addressing important issues with youth such as end of life or disability are present when it comes to patient-doctor-communication. Systemic barriers can be found, when looking for standardized transition processes. Agreements on a transition policy usually are missing. The lack of social workers or care managers are additional factors that hamper successful transition (Lemly et al., 2013).

When stating systemic barriers in transition, we should not forget to mention barriers such as no payment for transition activities and lack of funding for research (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011; White & Cooley, 2018). Rheumatologists in Brazil, resurveyed barriers to transition compared to the situation in US and Canada. They found a situation quite similar regarding the need of transitional care but due to financial pressure transition related issues were not prioritized in Brazil (Anelli, Len, Terreri, Russo, & Reiff, 2018). This fact might be true for other countries or geographical regions as well, it is definitely true for the situation in German speaking countries ("Deutsche Gesellschaft für Transitionsmedizin," 2018). A value based transition payment such as payment strategies, payment options, fee-for-service payments, has been recommended internationally (McManus, White, & Schmidt, 2018) in order to overcome some of the systemic barriers.

Additional findings point out to include ecological parameters such as geographic area, median income, sex composition or spoken language as risk factors on transitional outcomes (Javalkar et al., 2016).

1.3.3. Guidelines for HCT

Within the last years, an increasing number of position papers, consensus statements and guidelines for transitional care have been published for a considerable number of chronic conditions, eg. for renal diseases (Watson et al., 2011), for patients after transplant (Webb et al., 2010), for coeliac diseases (Ludvigsson et al., 2016), for TS (Bondy, 2007) or rare illnesses such as SCD (Bryant et al., 2014) kidney diseases (Andreoni et al., 2013; M. Ferris et al., 2015), rheumatic conditions (Felsenstein, Reiff, & Ramanathan, 2015), CF (Tuchman et al., 2010) and for type 1 diabetes (A. Peters, Laffel, & the American Diabetes Association Transitions Working Group, 2011).

In addition, comprehensive guidelines describe basic tenets crucial for transition. In 2011, the American Academy of Pediatrics (AAP) (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011) submitted an algorithm for the HCT process, addressing the need of pediatric HCP to guide their adolescent patients through the transition phase. The authors stated that transition planning should be an integrated part of routine medical care for all AYA. Therefore, individualized HCT services should be delivered to YSHCN as a matter of course in four steps:

- discussing the current transition policy,
- starting a transition plan,
- evaluating the plan at a certain time point and
- establishing an adult care model

In 2018, an updated version of the clinical report from 2011 extends considerations and recommendations with regard to infrastructure, payment, research and education (White & Cooley, 2018).

Furthermore, the National Institute for Health and Care Excellence (National Institute for Health and Care Excellence (NICE)., 2016) published a comprehensive guideline comprising more than 200 pages in 2016, presenting nine key elements:

- Age-banded clinics
- Meeting of adult team before transfer
- Promotion of health self-efficacy

- Written transition plan
- Appropriate parental involvement
- Key transition worker for each person
- Coordinated healthcare team
- Holistic life skills training
- Transition manager for clinical team

Notwithstanding all these efforts, gaps in the transitional care still persist. The implementation of structured tools has not been adopted nationwide and education of pediatric providers for adequate transition is still lacking (Nakhla, Bell, Wafa, & Dasgupta, 2017).

1.4. Overview on established transition programs

Meanwhile disease specific transition programs comprising generic aspects such as validation tools, structured questionnaires or assessment tools are on their way to be established in routine clinical care (Rieger et al., 2019). Starting the transitional process at an early age, these initiatives are keen to follow recommendations from the literature.

1.4.1. Gottransition™

The Gottransition program ("GotTransition.org," n.d.) is a joint initiative between the Maternal and Child Health Bureau and The National Alliance to Advance Adolescent Health situated in Washington, DC, USA. The main objective is to improve transition through new findings and innovative strategies. Further, the Gottransition™ expert group acts as a political driver, elaborating position papers as well as recommendations, for instance on transition payment fees. The process of HCT is illustrated on the basis of six core elements: developing a transition policy, tracking and monitoring transition, assessing transition readiness, planning transition, carrying out transfer of care, evaluating completion of transition. The six core elements intent to acquaint health care providers as well as youth and their caregivers with transitional issues. The written information serves as a guideline for a step-by-step implementation for transition services. Gottransition™ should rather be considered as a conceptual frame of transition activities than a specific transition program.

Figure 1: Website got transition

Download from https://www.gottransition.org/index.cfm, May 11, 2019

1.4.2. "Ready Steady Go"

The generic transition program "Ready Steady Go" (Nagra, McGinnity, Davis, & Salmon, 2015) is established as part of routine medical care in a large National Health Service teaching hospital in the UK. The key priority of the program is the empowerment of YSHCN. Questionnaires guide AYA and parents through topics relevant to successfully navigate the health care systems. The ready steady go-approach uses a traffic light system, where all documents related to the "ready"-state are linked to the color red, the "steady"-state are linked to the color orange and documents related to the "go"-state are in green color. Patient information leaflets and questionnaires are provided for downloads. Recommendations for age appropriate transition activities share the same color scheme.

The implementation of the generic program for patients with CF resulted in better prepared AYA and led to a more smoothly transfer to adult services (Connett & Nagra, 2018).

1.4.3. On our own feet ahead!

The Dutch transition program "On Your Own Feet Ahead" (van Staa, van der Stege, Jedeloo, Moll, & Hilberink, 2011) deals with identified bottlenecks in the process of transfer of YSHCN from pediatric centers to the adult medical home. A longitudinal study embedded in the transition program "On Your Own Feet Ahead!" investigated primarily the perception of the health provider team and AYA regarding team climate and quality of chronic illness care at two time points. Within a 1-year period improvements became evident (Cramm, Strating, & Nieboer, 2014; Nieboer et al., 2014).

1.4.4. Berliner Transitionsmodell (BTM)

The BTM (K. Minden, Niewerth, & Müther, 2014; Silvia Müther & Findorff, 2016) is a transition program developed by care providers working in the rheumatology department at the DRK Kliniken Berlin. However, the BTM is applicable for chronic conditions other than rheumatic conditions. Core element is a structured two-year

transition plan that actively involves patients, parents and care providers. A case manager acts as a continuous contact person throughout the transition process. He or she collects relevant documents such as the epicrisis, coordinates consultation hours jointly by a pediatrician and by an adult health care provider and assesses individual needs of the patients. Additionally, specific procedure documentations were developed for the BTM. Topics addressed in the BTM are not exclusively medical issues such as disease management and recommended follow-ups but also topics such as transition readiness, social integration, vocational training and plans-for-the-future. Leaflets with information for AYA, parents, resident doctors and health experts are available for downloads ("Www.btp-ev.de," 2019). The BTM is the first structured transition program in Germany, at least at present financed by the health insurance funds.

Figure 2: Website BTM

: downloaded from ("Www.btp-ev.de," 2019) on June 21, 2019

1.4.5. ModuS

A generic approach constitutes the **ModuS** transition workshops ("Kompetenznetz Patientenschulung e.V. - ModuS Transitionsschulung," 2016) designed for YSHCN as well as for their families. Especially trained persons teach content relevant for the transfer to adult care in three training sessions, two for

adolescent patients and one for their caregivers. The overall goal of these workshops is to empower YSHCN and sensitize parents. In addition to information about the health system and legal regulations, essential points are vocational training, career options and social life. Innovative didactic methods actively involving participants, role models and the use of new media complement the interactive workshops. Supplementary, a website www.between.kompas.de ("Www.between.kompas.de," 2016) provides well-prepared and detailed content consistent with the workshops. An accompanying study evaluating 45 workshops at 33 study sites showed higher transition related knowledge in former transition workshop participants and less parent involvement when it comes to disease management compared to the control group. No differences were observed regarding QoL. Overall, the workshop offer was well-received and rated as helpful. Nevertheless, the funding of the workshops still remains a major issue and has not entirely been solved (Menrath et al., 2018). ModuS workshops, however, are currently part of the BTM program ("Www.btp-ev.de," 2019)

Figure 3: Website between-kompas

1.4.6. NTx360

The project ("NTx360," 2019) is an innovative approach to secure long-term care for AYA after kindney transplantation in Germany. Aim of the initiative is optimal long-term care for all patients. Developed at the Hannover Medical School, the hospital-overlapping project is multimodal and based on telemedicine. Key elements include an electronic medical record and joint meetings of hospital medical doctors, resident medical doctors together with psychologists, physical therapists and case managers. Patient and care providers can access the electronic patient file. Video conferences as well as internet-based telemedical services complete the offering of NTx360.

Figure 4: Website NTx360

Downloaded from https://ntx360grad.de/ on July 13, 2019

1.5. Recommendations for HCT preparation

As guidelines for transitional care have been established, an increasing number of recommendations for the HCT preparation has become available. Nevertheless, suggestions have different priorities. The focus is either patient-orientated, system-orientated or process-orientated approach. Examples of patient centered recommendations include suggestions how to approach transition successfully in terms of transition preparation activities in the clinical setting. Demands for a standardized, timely initiated transition processes in a multi-disciplinary setting are raised in most comprehensive guidelines (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011; White & Cooley, 2018) .

Descriptions on how to facilitate health care self-management (M. Ferris et al., 2015) and how to emphasize the importance of adolescents' perspective (Fredericks, 2017) are available. A fundamental difference in approaching HCT is the setting in which activities shall take place.

Recommendations for the pediatric setting and for the adult care setting differ. Within the pediatric setting, timely addressing transition issues with adolescents and caregivers (Bryant et al., 2014), eventually starting at the age of eleven (Nagra et al., 2015), and introducing private and confidential discussions with AYA (M. Ferris et al., 2015) is recommended. Psychosocial issues need to be identified and addressed (Iyengar, 2019) as well as ecological factors that may facilitate HCT (M. Ferris et al., 2015). A major reluctance of AYA and their families to leave their pediatric provider and the familiar hospital staff may impede transition activities (Lemly et al., 2013). Cooperation with the adult care team is strongly recommended (Lemly et al., 2013; Bryant et al., 2014; M. Ferris et al., 2015; White & Cooley, 2018) and might decrease the reluctance. Additionally, annual assessment of transition readiness or self-management skills (Sawicki et al., 2011; Lemly et al., 2013; Bryant et al., 2014; M. Ferris et al., 2015) as well as other objective assessments (Iyengar, 2019) are recommended. Reinforced patient education (A. Peters et al., 2011; White & Cooley, 2018) to optimize self-management (Calhoun, Abel, Pham, Thompson, & King, 2019) and empowerment of YSHCN (Thyen et al., 2016) are regarded as helpful. Repeated discussions on the fact of transfer and related questions, such as education, social

perspectives as well as the topic of intimate relationships (Calhoun et al., 2019) should be matter of fact (van Staa et al., 2011).

A model for effective receivership has been introduced, comprising five recommendations for the adult setting. These recommendations include the emphasis on a team-based approach encompassing the cooperation with pediatric colleagues and the focus on a good relationship with the young patient instead to focus on blood glucose levels (Iyengar, 2019).

The idea of a professional transition manager or “transition coordinator”, who coordinates telephone or email contact and who schedules joint appointments with pediatricians and adult care providers was presented in the world of diabetes patients years ago (Weissberg-Benchell, Wolpert, & Anderson, 2007). Providing information via a designated website and a written summary on access to adult providers, insurance coverage and therapy regimen have been recommended equally to assessment of YSHCN´s knowledge and skills regarding their diagnosis.

Australian authors stated already in 2008 that transition issues such as research, training and hospital services need to be addressed in overarching collaboration (Kennedy & Sawyer, 2008). A credentialing process for providers and for standards of care (American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group, 2011) as well as securing continuity of care (Sheehan et al., 2015) was argued years ago.

1.6. Recommendations for structured assessment of HCT readiness

Clinicians as well as literature call for more autonomy in AYA when it comes to transitional issues (Rosen et al., 2003). However, autonomy is not a linear concept. Health autonomy is comprised of sophisticated skills which can be subdivided into attitudinal, emotional and functional autonomy (Noom, Deković, & Meeus, 2001). Moreover in the healthcare context, autonomy is often put on a level with self-regulation, motivation and health-related behavior (Ryan, 2008). In addition, competencies such as self-reliance, awareness of responsibilities for therapy regimen and medication adherence as well as skills in self-organization are often subsumed under the term “autonomy” (Kennedy & Sawyer, 2008). Autonomy in specific domains can appear jointly with a lack of autonomy in other areas (Sattoe, Hilberink, van Staa, & Bal, 2014). The effort to increase wellbeing and decrease paternalization for patients with cognitive impairments or mental health problems is necessary. However, limitations in research ground in a small evidence base. There are no or very few reports on sampling strategies and a lack of data saturation is evident (the MILESTONE Consortium et al., 2018).

Out of nearly 1000 AYA in a Dutch university hospital, who participated in a web-based study and filled out a questionnaire on transition readiness, only about 50% felt ready for transition. Associated factors to self-perceived transition readiness were, in line with other studies, older age, but also a positive attitude towards transition and having had discussions related to transfer issues. The highest readiness for transition were found for AYA who reported autonomously visiting their medical doctors or other hospital appointments and who regarded themselves more independent in the consultation setting (van Staa et al., 2011). However, discrepancies in the perception of disease management responsibilities and self-care skills between adolescents and parents or providers are common (Fredericks, 2017).

Adequate care for developmental age of YSHCN is acknowledged to be essential in transition services (White & Cooley, 2018). Therefore, recommendations for improved preparation for transition favor a timely assessment of transition readiness instead of simply transferring YSHCN from pediatric care to adult health care by the age of eighteen (Fair et al., 2015; Sawicki et al., 2011; Zhang-Jiang & Gorter, 2018).

Determination of transition readiness is now seen as a crucial element that effects positively the patient's outcome (Zhou et al., 2016). HCT readiness assessment is important when it comes to shifting disease related responsibilities from caregivers to AYA. Perceived burden of responsibilities or self-rated management skills might be overrated by AYAs, whereas their parents might underestimate the disease related burden (van Staa et al., 2011). Recommendations advise to first assess transition readiness and in a next step to decide on when and how responsibility will be transmitted, accordingly to the assessment´s results (Kaugars, Kichler, & Alemzadeh, 2011). Recently, discussions on assessing transition readiness in patients with rare diseases are more and more common (Calhoun et al., 2019).

1.7. Transition Readiness Assessment Tools

There are hardly any well-studied, validated and therefore established HCT assessment tools in German language. This fact currently hampers the process of HCT readiness assessment in German speaking countries. Internationally, the need for a valid and reliable transition readiness assessment instrument was claimed more than a decade ago (Kennedy & Sawyer, 2008). A systematic review dated 2014 investigated studies on transfer satisfaction measurements as well as on assessment of transition readiness tools and stated a major gap in knowledge and availability of well-studied and validated readiness assessment tools (Stinson et al., 2014). An example for misuse of non-validated questionnaires is the application of the Rotterdam Profile (Donkervoort et al., 2009) It was originally designed to describe the transition process of AYA with cerebral palsy (CP). A review after ten years of application of the Rotterdam profile showed that the instrument was used as a transition measurement instrument in all kinds of chronic conditions. However, no psychometric analyses were conducted (Zhang-Jiang & Gorter, 2018).

Another systematic review in 2014 identified ten different HCT assessment tools in English language (Zhang et al., 2014). Out of ten, only three tools were generic: the UNC TRxANSITION (M. E. Ferris et al., 2012), the Self-Management Skills Assessment Guide (Williams, Dunseith, & Mah, 2010) and the TRAQ (Sawicki et al., 2011; Wood et al., 2014). Solely, the TRAQ demonstrated adequate psychometric measurement qualities with sufficient content validity, construct validity and good internal consistency.

1.7.1. The UNC TR$_x$ansition Scale

At the University of North Carolina, the TR$_x$ansition Scale (M. E. Ferris et al., 2012) was developed to measure health-care transition and self-management skills of YSHCN, to be administered in clinical settings. Theoretical models, literature search, expert opinion and feedback from YSHCN and their caregivers led to the 33-item semi-structured interview comprising 10 domains. The UNC TR$_x$ansition Scale is not solely based on self-report but is also validated by information from the medical records. The

scale was administered in the public health sector (Javalkar et al., 2016) and was cross-culturally adapted to Malayan (Chong, 2018).

1.7.2. The STARx Development

The STARx Questionnaire (Cohen et al., 2015; M. Ferris et al., 2015) was designed as a generic assessment tool, not dedicated to a single chronic condition but for use across various childhood onset chronic conditions. It is a self-report instrument, based on a six-factor structure (M. Ferris et al., 2015) addressing the following thematic areas: medication management, provider communication, engagement during appointments, disease knowledge, adult health responsibilities, resource utilization. A subsequent validation of the STARx tool (Cohen et al., 2015) showed good concurrent validity, correlated with the UNC Tr$_x$ansition scale (M. E. Ferris et al., 2012) and the TRAQ 5.0 (Wood et al., 2014). Moreover, higher self-reported transition readiness was significantly associated with higher assessed transition readiness by the patient´s care providers. When correlating the tool with validated questionnaires assessing health literacy, self-efficacy (Iannotti, Schneider, Nansel, & Haynie, 2006) or medication adherence, the STARx proved good predictive validity. However, the instrument showed no discriminant validity when looking for significant differences among three different conditions (kidney disease, lung disease, gastrointestinal disease) with regard to the STARx scores. The authors pointed out that the process of transition might rather be related to skills within an individual than be associated with a specific diagnosis (M. Ferris et al., 2015).

1.7.3. Self-Management Skills Assessment Guide

The questionnaire (Williams et al., 2010) comprises 21 items, rated by YSHCN and parents. Only a small number of participants (n=49) were included in the pilot study. Items are identically structured. They mainly address self-management skills. Cronbach´s alpha proved to be good.

1.7.4. Transition Readiness Assessment Questionnaire (TRAQ)

The American TRAQ was developed to prepare adolescent patients for successful transition to adult medical care. The initial development took place at the University of Florida, Jacksonville. An extensive research comprising literature search, comparison of transition related web sites as well as a comprehensive analysis of several transition programs led to the conclusion that no validated tool for use in the transition process existed. Consequently, the resulting 33-items questionnaire was fielded to 192 young patients at two sites. The authors regarded all items as skills and, therefore, the original response set was based on the transtheoretical approach and the model of change (Prochaska & DiClemente, 2005). Psychometric analysis comprised Exploratory Factor Analysis (EFA), assessment of Cronbach´s alpha and known-groups validity for age, gender, race and primary diagnosis.

This work resulted in the first version of the TRAQ comprising 29 items, divided in two domains. Domain 1 primarily assessed skills for self-management, whereas domain 2 assessed self-advocacy competencies (Sawicki et al., 2011). Further development of the TRAQ and additional psychometric analyses led to the actual TRAQ 5.0 (see appendix) with 5 subscales and 20-items (Wood et al., 2014). The TRAQ 5.0 is a validated, self-reported and disease neutral questionnaire in English language. It was designed for assessing self-management skills as well as self-advocacy competency and therapy adherence in YSHCN.

A systematic review (Zhang et al., 2014) of psychometric properties attested the TRAQ (Sawicki et al., 2011; Wood et al., 2014) to be the best validated tool with high internal consistency and good construct validity as well as adequate content validity. Additional benefit was seen in the TRAQ´s generic character.

Figure 5: Transition Readiness Assessment Questionnaire, TRAQ 5.0 (Wood et al., 2014)

Patient Name: _____ **Date of Birth:** ___/___/___ **Today's Date** ___/___/___ **(MRN#** _____**)**

Transition Readiness Assessment Questionnaire (TRAQ)

Directions to Youth and Young Adults: Please check the box that best describes ***your*** skill level in the following areas that are important for transition to adult health care. There is no right or wrong answer and your answers will remain confidential and private.

Directions to Caregivers/Parents: If your youth or young adult is unable to complete the tasks below on their own, please check the box that best describes ***your*** skill level. **Check here** if you are a parent/caregiver completing this form. ☐

	No, I do not know how	No, but I want to learn	No, but I am learning to do this	Yes, I have started doing this	Yes, I always do this when I need to
Managing Medications					
1. Do you fill a prescription if you need to?					
2. Do you know what to do if you are having a bad reaction to your medications?					
3. Do you take medications correctly and on your own?					
4. Do you reorder medications before they run out?					
Appointment Keeping					
5. Do you call the doctor's office to make an appointment?					
6. Do you follow-up on any referral for tests, check-ups or labs?					
7. Do you arrange for your ride to medical appointments?					
8. Do you call the doctor about unusual changes in your health (For example: Allergic reactions)?					
9. Do you apply for health insurance if you lose your current coverage?					
10. Do you know what your health insurance covers?					
11. Do you manage your money & budget household expenses (For example: use checking/debit card)?					
Tracking Health Issues					
12. Do you fill out the medical history form, including a list of your allergies?					
13. Do you keep a calendar or list of medical and other appointments?					
14. Do you make a list of questions before the doctor's visit?					
15. Do you get financial help with school or work?					
Talking with Providers					
16. Do you tell the doctor or nurse what you are feeling?					
17. Do you answer questions that are asked by the doctor, nurse, or clinic staff?					
Managing Daily Activities					
18. Do you help plan or prepare meals/food?					
19. Do you keep home/room clean or clean-up after meals?					
20. Do you use neighborhood stores and services (For example: Grocery stores and pharmacy stores)?					

1.7.5. Cross-cultural adaptation and clinical use of the TRAQ

Since the TRAQ proved to be a useful instrument with good psychometric properties, clinicians from around the world became interested in the tool. So far, the TRAQ has been translated and cross-culturally adapted into Argentinian Spanish (DeCunto et al., 2017; Gonzalez et al., 2017), into Brazilian Portuguese (Anelli et al., 2018) and into Turkish (Kızıler, Yıldız, & Eren Fidancı, 2018a).

The different versions of the TRAQ questionnaire were used in multifaceted studies over the last years. At the San Francisco Cystic Fibrosis Center, a transition program for patients with CF used a standardized transition protocol and embedded in the curriculum the use of the TRAQ. At follow-up eighteen months later, transition outcomes improved significantly (Okumura et al., 2014).

Studies report higher TRAQ scores associated with older age (Sawicki et al., 2011; Wood et al., 2014; Stewart et al., 2017; Anelli et al., 2018). Nevertheless, there are other parameters than age that account for transition readiness as measured by the TRAQ. Some studies indicate that gender accounts for differences in TRAQ scores (Sawicki et al., 2011), others did not find gender related differences (Jensen et al., 2017). Health care transition skills according to TRAQ scores were associated with self-care beliefs (Sawicki et al., 2014). In AYA with IBD, self-efficacy and resilience predicted transition readiness scores measured with the TRAQ questionnaire (Carlsen et al., 2017).

Intentional self-regulation and hopeful expectations for the future positively correlated with higher TRAQ scores and, therefore, might account for transition readiness (Hart, Pollock, Hill, & Maslow, 2016). Similarly, when patients were asked for their future perspectives, it proved that patients with “plans for the future” scored higher than patients without plans (Gonzalez et al., 2017).

Other findings suggested that parental involvement in the HCT process and higher parental medical condition knowledge is associated with increased transition readiness in YSHCN (On behalf of the Transition Collaborative Group et al., 2018; Stewart et al., 2017). This goes in line with earlier research indicating that parents’ and teens’ health literacy is entangled in the context of HCT (Chisolm, Sarkar, Kelleher, & Sanders, 2015).

Furthermore, patient and family characteristic as well as condition accounted for differences in TRAQ scores. Disparities among different chronic conditions regarding transition readiness were found on an item-level (Beal et al., 2016). Analysis indicated that appointment-keeping, activities of daily living and medication accounted for significant variances. Youth with autism spectrum disorder (ASD), a diagnosis comprising impairments in the neurocognitive area, scored significantly lower than youth with chronic conditions without impediments in cognition and social behavior, such as type 1 diabetes or rheumatic conditions (Beal et al., 2016).

1.8. Challenges for HCT for different chronic conditions

A paucity of effective transitional processes exists for patients diagnosed with a rare disease, despite the acknowledged need of lifelong health care. Insufficient medical care in adult patients with persistent GHD (Courtillot et al., 2013) and patients with SCD (Bryant et al., 2014) are common. In line, failed transition can be stated for women with TS (Ertl et al., 2018). According to follow-up studies regarding transition and data on adequate medical care, there seem to be very different dynamics in the various chronic conditions. Care pathways seem differently foretold with more robust pathways for more common chronic diseases compared to rare diseases.

The question whether patients with complex chronic conditions face additional impediments with a view on long-term medical outcome has been answered by the literature (Bachelot et al., 2017; Courtillot et al., 2013; Ertl et al., 2018).In addition, decreased HCT readiness scores for conditions associated with cognitive impairments were reported (Sawicki et al., 2011; Beal et al., 2016). Cognitive functioning is a crucial parameter for the developmental trajectories in children and adolescents and therefore a critical factor in the process of transitioning (Beal et al., 2016).

1.8.1. Turner syndrome (TS) and HCT

TS falls within the group of rare diseases, with an incidence of 1:2500 in female births (Ford, Jones, Polani, De Almeida, & Briggs, 1959; Hall & Gilchrist, 1990). Typical clinical pictures seen in TS comprise short stature (Haeusler et al., 1992; Haeusler & Frisch, 1994; Haeusler et al., 1995, 1996a; Haeusler, 1998), ovarian dysgenesis, structural heart and/or renal defects, together with other possible phenotypic characteristics such as metabolic malfunction, autoimmune defficiencies or an increased risk for hearing impairments (P. Saenger et al., 2001; Bondy, 2007; Mortensen et al., 2009; Davenport, 2010; Backeljauw et al., 2015; Schmitt, Haeusler, Blümel, Plöchl, & Frisch, 1997; Haeusler et al., 1996b). These multiple health issues result either from structural anomalies in the X chromosome or the complete loss of it (45X) (Ford et al., 1959; Hall & Gilchrist, 1990). Furthermore, TS is a condition that includes a broad range of psychosocial challenges (David S. Hong, Dunkin, & Reiss, 2011; McCauley, Feuillan, Kushner, & Ross, 2001) as learning difficulties (J. L. Ross et

al., 2002; Judith L. Ross, Roeltgen, Kushner, Wei, & Zinn, 2000; M. T. Ross et al., 2005; D. S. Hong & Reiss, 2012), decreased arithmetic abilities (Baker & Reiss, 2016; M. M. M. Mazzocco, 2006), impairments in executive functioning and planning skills (D. Hong, Scaletta Kent, & Kesler, 2009; Kirk, Mazzocco, & Kover, 2005) as well as autism spectrum traits, attention deficiency and hyperactivity disorder (ADHD) (Russell, 2006). Whereas verbal skills count to the specific strengths of TS (M. M. Mazzocco et al., 2006; Temple, 2002; Temple, cristine; Shepard, Elizabeth, 2012).

In addition, pubertal development is either delayed or menstruation and growth of female breasts stay away as a result of failed ovarian function and estrogen deficit (Bondy, 2007; Bondy et al., 2007). This fact makes pubertal and fertility issues even more challenging (Grynberg et al., 2016; Sheaffer, Lange, & Bondy, 2008) than for other adolescents with chronic conditions not affecting sexual development. Lower self-esteem (Carel et al., 2006) and emotional immaturity (McCauley et al., 2001; Sybert & McCauley, 2004) add to the complex situation and challenging health care demands of adolescent girls with TS (Pinsker, 2012).

Facing a condition becoming more and more complex with age, adult women diagnosed with TS need life-long medical surveillance (Devernay, Ecosse, Coste, & Carel, 2009; Downing et al., 2013). Under 4% of adult women with TS transitioned successfully, in the sense of adequate medical care in adulthood (Devernay et al., 2009; Ertl et al., 2018). Since medical doctors familiar with TS in adulthood are hard to find, women with TS need to have full awareness of their condition (Davies, 2010), Yet, young women with TS often do not show full understanding of their diagnosis (Stalla et al., 2013). Studies indicate that deficits in medical treatment for patients are ubiquitous (Gawlik et al., 2012; Gawlik & Malecka-Tendera, 2014), as women with TS do not see specialized doctors and do not undergo recommended follow-ups (Ertl et al., 2018). For TS, clinical guidelines for structured medical follow-up in adulthood do exist, however, there has no gold standard for structured transition been established yet (Paul Saenger, 2004; Bondy, 2007; Gleeson, Davis, Jones, O'Shea, & Clayton, 2013; Downing et al., 2013; Backeljauw et al., 2015). Nevertheless, an intensified focus can be noted and an updated review on transitional care for girls with TS is available (Bernard et al., 2019).

1.8.2. Type 1 diabetes mellitus and HCT

Type 1 diabetes mellitus is a complex metabolic disorder and is characterized by chronic hyperglycemia resulting from defects in insulin action or insulin secretion (Danne et al., 2004; Rami-Merhar, Fröhlich-Reiterer, & Hofer, 2016). The diagnosis of type 1 diabetes demands lifelong insulin substitution and requires measuring blood glucose levels several times per day (and night), control over food intake and records on blood measurements and administered doses of insulin (Kordonouri et al., 2014; Mayer-Davis et al., 2018). Worldwide, type 1 diabetes is the most common metabolic disorder in youth with an incidence significantly varying between different countries and regions (Rogers, Rogers, & Basu, 2018), with an incidence of 0,34/100.000/Y in Austria (Rami-Merhar et al., 2016).

In the lives of AYA with type 1 diabetes the transitional phase is associated with poor adherence to insulin regimen, inadequate blood sugar control and increased blood sugar levels as well as risk for acute complications and hospitalization (Cameron et al., 2014; für das Kompetenznetz Diabetes et al., 2009; Kipps et al., 2002). In children and adolescents diagnosed with diabetes, school achievements seemed to be stronger related to socioeconomic status and behavioral problems rated by parents than to metabolic control (McCarthy, Lindgren, Mengeling, Tsalikian, & Engvall, 2003). In contrast, a meta-analysis from 2008 found lower, however only moderately lower, cognitive performance in young patients with early onset disease, particularly for memory and learning skills (Gaudieri, Chen, Greer, & Holmes, 2008). Experience of severe hypoglycemia especially in young children seems to have long-term adverse effects on verbal skills or problem solving (Asvold, Sand, Hestad, & Bjørgaas, 2010)

A systematic review investigated the impact of HCT on diabetes outcome (Sheehan et al., 2015). Two types of studies were identified. On the one hand reports that focused on diabetes related outcomes such as glycemic control, attendance at clinics and at care providers or diabetes related admissions and on the other hand reports that described the experience of transition, practical and psychosocial issues including discontinuity of care. Though a scantiness of high-quality studies was stated, transition for AYA with type 1 diabetes seems to be associated with missed clinical

appointments whereas structured transition programs seem to have some positive impact. However, guidelines recommend transfer based on the patient´s readiness for transition (A. Peters et al., 2011; L. W. H. Peters et al., 2014).

1.8.3. Childhood onset rheumatic conditions and HCT

Juvenile idiopathic arthritis (JIA) is the term used for a group of rheumatic childhood onset conditions, formerly also known as juvenile chronic arthritis and juvenile rheumatoid arthritis. JIA is the most common childhood onset chronic systemic autoimmune disease (Günther & Trauzeddel, 2013). Therefore, the common method to treat JIA is using drugs that modulate the immune system (Dueckers et al., 2012). Although spontaneous remission is experienced from around two thirds of the children, one in three patients will have to live with life-long disease activity (Foster, Marshall, Myers, Dunkley, & Griffiths, 2003; Klotsche et al., 2016). In this case, AYA are in the need of specialized long-term follow-up. Rheumatic symptoms and associated co-morbidity persists into adulthood (Kirsten Minden et al., 2002).

Moreover, psychosocial burden appears to also persist into adult life. Research, in regard to long-term outcome in adult patients with JIA, indicated lower HRQoL and significant lower employment rates compared to controls, although the educational attainment was in the normal range compared to the local population group (Foster et al., 2003). Other studies underpin these findings, as cognitive functioning in AYA diagnosed with childhood onset arthritis showed normal range of intelligence quotient learning skills, working memory and attention. Motor skills are also within the normal spectrum, no social or emotional deficits were apparent (Feldmann, Weglage, Roth, Foell, & Frosch, 2005). Self-reported social functioning and scores in depression, anxiety, other emotional problems or relations to peers and behavior were not different from the norm population and not associated with disease activity (Ding, Hall, Jacobs, & David, 2008). A review (Dick & Riddell, 2010) on studies which investigated the association between cognitive performance and chronic pain in children with arthritis found that general intelligence was not affected by the condition and the fact of bearing up against pain. Transition, comprising early preparation of AYA and standardized,

well-documented processes has been an issue of discussions and been in the focus of pediatric arthritis experts for some years now (Ganser & Müther, 2012; K. Minden & Niewerth, 2012; S. Müther, Rodeck, Wurst, & Nolting, 2014).

Recommendations for transitioning in a wise and coordinated way do exist on behalf of the British Society of Paediatric and Adolescent Rheumatology, where the impact of a coordinated transitional care program on adolescents with juvenile idiopathic arthritis is described (McDonagh, Southwood, Shaw, & on behalf of the British Society of Paediatric and Adolescent Rheumatology, 2007). In the course of HCT of JIA patients, assessment of transition readiness is suggested (White & Ardoin, 2015).

1.9. Aims of the thesis

The transition phase from pediatrics to adult medical care still is a critical stage in the lives of adolescents with chronic conditions. Standardized procedures are often lacking and procedures are unclear. A growing body of evidence and recommendations from the literature emphasize assessing transition readiness. To date, no disease neutral questionnaire in German language exists to assess skills crucial for adolescents with chronic medical conditions.

In daily clinical practice, feasible questionnaires that can be administered time efficiently and interpreted easily are a necessity. In the present study, I primarily aimed at providing an easy-to-use screening tool for assessing transition readiness for YSHCN in German speaking countries. Secondly, I wanted to test the questionnaire´s benefit for clinical research. I was especially interested in potential differences in transition readiness between patients with TS and more common chronic medical conditions to better understand our patients' skills and deficits in managing their disease before being transferred to adult services.

Objective 1: to translate and cross-culturally adapt the TRAQ 5.0 and to develop a German version (GV) of the TRAQ

Objective 2: to pilot-test the TRAQ-GV with a heterogeneous study cohort of YSHCN aged 14 to 23

Objective 3: to conduct descriptive statistics and psychometric analyses

Objective 4: to assess feasibility and time efficiency of the administration of TRAQ-GV

Objective 5: to test the administration of the TRAQ-GV in a patient group with TS and to investigate differences in transition readiness according to TRAQ-GV scores between adolescents with TS and a control group comprising female adolescents diagnosed with either type 1 diabetes or a rheumatic condition

Anticipated benefits for patients

Direct benefit for our adolescent patients will arise in the course of questionnaire administration from addressing issues related to the transitional process. YSHCN, parents and caregivers as well as health care providers will be sensitized for the demands while transitioning and will eventually start to discuss the process of transition in a support oriented way, including issues such as disease knowledge, therapy regimen, medication, self-management skills and self-advocacy competency.

Anticipated clinical benefits

The adaptation of a disease neutral transition readiness questionnaire will possibly lead to the implementation of standardized assessment within the transition process in the routine clinical care. The newly adapted tool will provide a baseline measurement for a relevant number of YSHCN at our clinic, follow-ups in the course of the transition phase will have comparative values.

Figure 6: Workflow overall

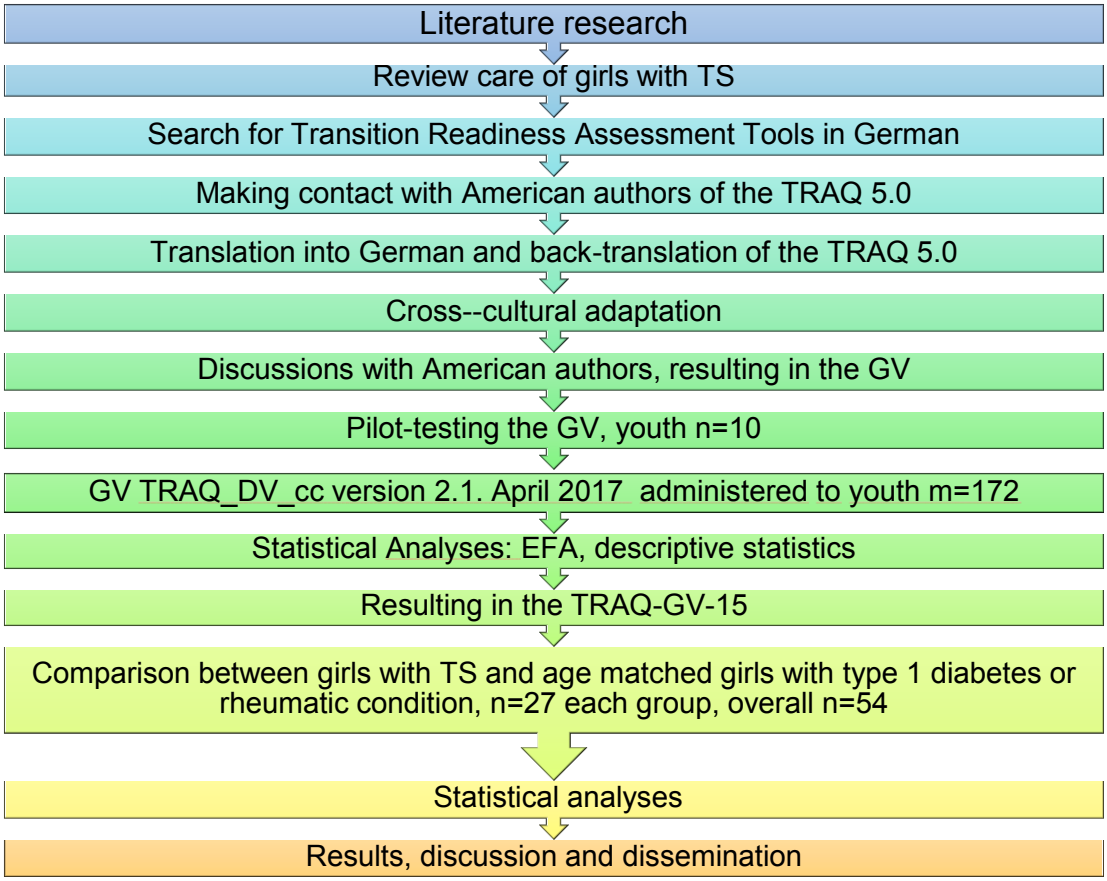
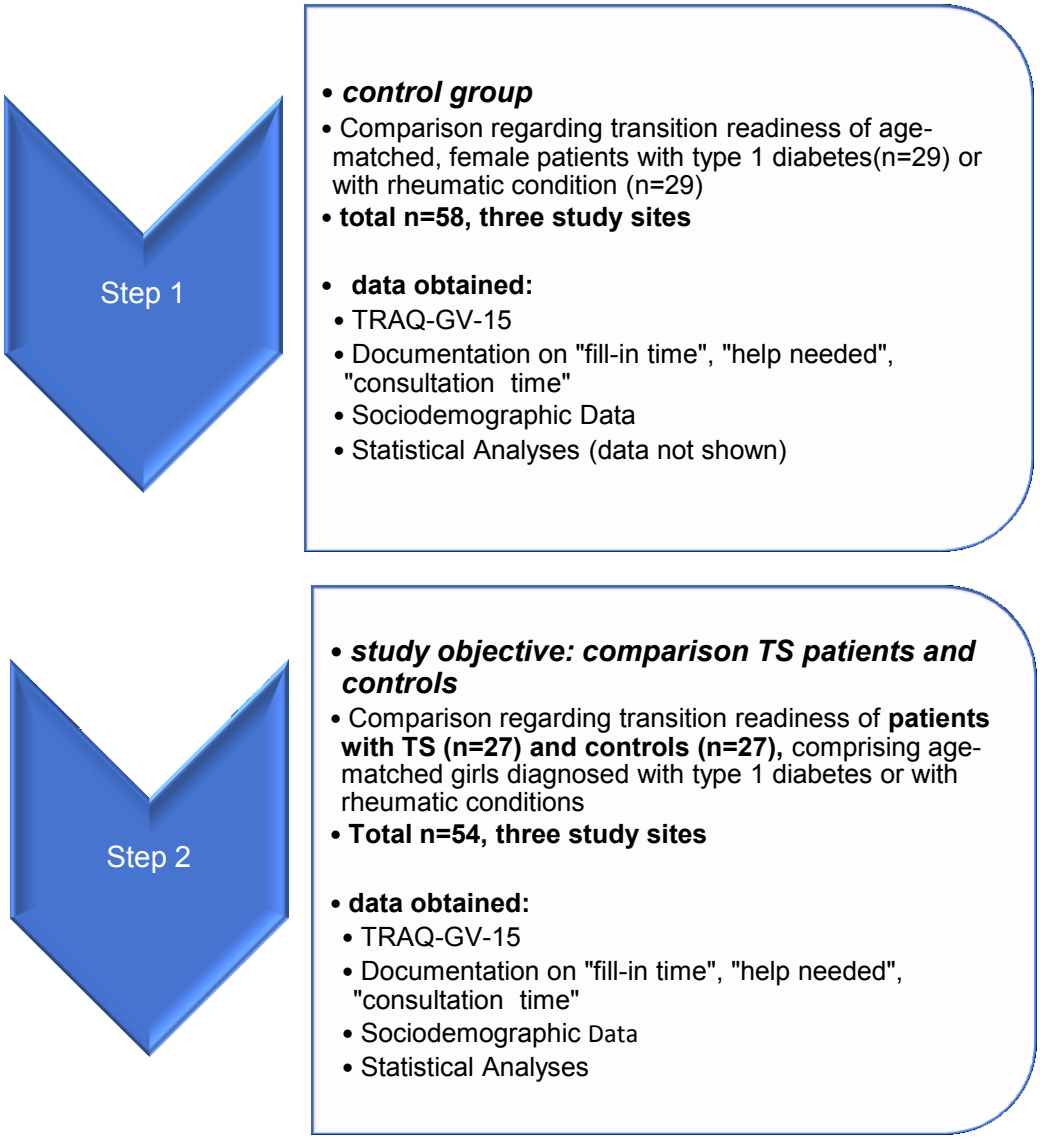


Figure 7: Workflow TS study



CHAPTER TWO: RESULTS

2.1. Care of girls and women with Turner syndrome: beyond growth and hormones.

Culen C, Ertl DA, Schubert K, Bartha-Doering L, Haeusler G.
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PMID: 28336768

Turner syndrome (TS) is a complex genetic disorder with a global incidence of 1 in 2500 female births, determined by loss or an anomaly of the X-chromosome. The TS phenotype includes short stature, delayed or omitted pubertal development and a risk for organ dysfunction. A multifaceted TS-specific neurocognitive profile may comprise deficits in executive functioning, learning difficulties, a lack of social skills, autism traits and a decreased autonomy. Treatment of TS includes hormonal substitution as well as treatment of clinical symptoms. Comprehensive and updated guidelines for medical treatment are available. Patients with TS need life-long medical follow-up. Vocational career, intimate relationships and family life remain a challenge for TS patients. However, psychological issues in girls and women were given too little attention in the past years. Therefore, I provided an updated review on care of girls and women with TS beyond growth hormone therapy and estrogen substitution, including recommendations on diagnosis disclosure, psychological testing and counseling for socio-psychological issues. Transition from comprehensive pediatric care to medical care providers in the adult setting is especially difficult for patients with TS.

My contribution to the review consisted in the creation and development of the concept, in reviewing the literature, in revising adequate psychological test instruments and in the manuscript preparation.

Review
Open Access

C Culen *et al.* | Turner syndrome: beyond growth and hormones | **R39**–R51 | **6**:R39

Care of girls and women with Turner syndrome: beyond growth and hormones

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Abstract

Turner syndrome (TS), although considered a rare disease, is the most common sex chromosome abnormality in women, with an incident of 1 in 2500 female births. TS is characterized by distinctive physical features such as short stature, ovarian dysgenesis, an increased risk for heart and renal defects as well as a specific cognitive and psychosocial phenotype. Given the complexity of the condition, patients face manifold difficulties which increase over the lifespan. Furthermore, failures during the transitional phase to adult care result in moderate health outcomes and decreased quality of life. Guidelines on the optimal screening procedures and medical treatment are easy to find. However, recommendations for the treatment of the incriminating psychosocial aspects in TS are scarce. In this work, we first reviewed the literature on the cognitive and psychosocial development of girls with TS compared with normal development, from disclosure to young adulthood, and then introduce a psychosocial approach to counseling and treating patients with TS, including recommendations for age-appropriate psychological diagnostics. With this work, we aim to facilitate the integration of emphasized psychosocial care in state-of-the-art treatment for girls and women with TS.

Key Words
- psychosocial care in endocrinology
- psychological approach
- x-linked
- development in Turner syndrome
- cognitive profile
- psychosocial recommendations in peadiatrics
- health autonomy
- transition in endocrinological care

Endocrine Connections (2017) **6**, R39–R51

Introduction

Turner syndrome (TS) is caused by structural anomalies in or complete loss of the X-chromosome (45X). Although a rare disease with an incidence of 1 in 2500 female births, it is nevertheless the most common sex chromosome abnormality of human females. TS is characterized by short stature and ovarian dysgenesis, together with a broad range of other phenotypic characteristics, including an increased risk for heart and renal defects (1, 2).

Clinicians treating girls with TS are challenged with many endocrine, genetic, cardiovascular, developmental, reproductive and psychosocial issues. Medical care of girls with TS in child-centered healthcare systems is therefore highly specialized (3, 4, 5, 6). Reviews of medical care in TS that propose clinical guidelines and recommendations occasionally also mention psychosocial issues (3, 4, 7, 8), but these are no more than footnotes compared with the medical discourse (9, 10).

Patients with TS have to cope with many difficulties; they not only need life-long medical surveillance but also psychological care and treatment for a good outcome (6). Symptoms emerge slowly. During the early infancy, affected girls show no psychological developmental differences from their unaffected counterparts. As they grow older their phenotypical, psychosocial and intellectual development is seen to more clearly diverge from that of their normally developing peers. The typical TS-specific cognitive profile is characterized by strengths in verbal skills (11) and relatively weak performance

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in arithmetic, visuospatial processing and executive functioning (12). In addition, girls with TS have frequently been found to have difficulty in encoding social cues and behaving appropriately in social situations (13). Some cognitive and psychosocial TS phenotypes such as deficits in mathematical abilities (14) and impaired social interaction (13) persist throughout the adult life.

Early diagnosis and treatment of comorbidities is known to enhance the medical state of adult patients with TS (15). Experienced clinicians have discussed inclusion of psychological testing and routinely screening of girls with TS for developmental progress, but evidence-based results are rare (16). Consequently, intervention guidelines for the psychosocial aspects of TS are scarce (13). To our knowledge, no evaluated and documented standardized best practice model is available to provide psychological support for girls and women with TS.

Transition, when adolescents move within the healthcare system from a pediatric protective and authoritative environment to an adult one that demands healthcare autonomy and a degree of independence and responsibility, is a difficult stage. Individual readiness for the process varies and cannot be assumed to be naturally completed by the age of 18 years (17). Previous studies have shown that many young women with TS are 'lost in transition' (18), suggesting that successful passage from pediatric to adult care requires a special focus in TS.

We have developed recommendations towards a psychological and psychosocial approach to improve the care for girls with TS. Our recommendations importantly include supportive interventions encompassing the family system and caregivers. Our overall aim is to empower patients with TS to keep up with their peers at school and in their working life, and to improve their self-esteem and thus their quality of life (QoL). Table 1 gives a summary of the relevant background information, recommendations and psychological tests.

Counseling and treating girls with TS is complicated because although the karyotype differences may affect the phenotype of TS, the karyotype is not a distinct predictor of the physical or psychological phenotypical outcome (19). Consequently, in TS we must acknowledge normal and healthy development while being attuned to the possible impairments.

We thus compare the development in TS with the expected norms of development (20, 21) and describe areas of potential impairment as well as possible interventions including psychological screening and testing.

Diagnostic disclosure

Clinical abnormalities upon which a diagnoses of TS can be made arise from prenatal life until adulthood (4, 22). These include suspicious findings of cystic hygroma, increased nucheal translucency in fetal ultrasound, lymphedema on hands and feet and cardiac malformations at birth, short stature during childhood, delayed puberty combined with short stature at adolescence and even premature ovarian failure during adulthood. Ultimately however, 20–30% of the girls are diagnosed immediately after birth because of phenotype characteristics such as puffy hands, edemas or a webbed neck (7).

Thus, the clinical setting or the practitioners charged with disclosing the TS diagnosis to the parents and/or the patient could be prenatal counseling geneticists, neonatal services, cardiologists, pediatricians, endocrinologists, gynecologists or other specialists. The questions from advice-seeking parents and patients who will require explanation can cover a wide range, from life-deciding aspects of prenatal counseling to short stature and infertility. In all situations, merely explaining that the girl has a chromosomal disorder responsible for a complex combination of multisystem symptoms of unpredictable severity is insufficient to meet many questions raised when a decision is made to investigate the karyotype.

Keeping the TS condition a secret has been shown to have an adverse effect on the patient's well-being, whereas full disclosure to the girl in an empathetic way has been found to support positive integration of the condition (23).

Recommendations

Given the complexity of a TS diagnosis and treatment, the diagnosis needs to be clearly communicated to the parents from the beginning, and to the child having regard to her age and developmental stage. The earlier the diagnosis is communicated, the better is the outcome for the girl's future QoL and self-esteem (23). Furthermore, honesty about the condition within the family is essential. Pediatricians should offer assistance to overstrained parents in disclosing the condition to their daughters in an age-appropriate manner. Providing written information on the most important issues of TS to facilitate digesting the diagnosis over a period of time might be worthy of consideration (23). The clinician charged with disclosing the diagnosis should be familiar with all aspects of TS. He or she should set aside sufficient time to inform the

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Table 1 Background information, recommendations and psychological tests.

Normative developmental concepts				
Age	Developmental tasks (Robert J Havighurst (20))	Cognition (Jean Piaget (21))	Turner syndrome: pullouts for psychosocial care	Psychological tools
Diagnosis disclosure			– Clear and empathic communication, in depth knowledge on the condition is required – Sufficient time to answer questions – Within the family: emphasizing being honest about diagnosis, full disclosure to the girl – Thorough documentation about information provided – Information about self-help groups, literature, etc. – Diagnosis disclosure is an ongoing process along the way of the girl's development	
Infancy 0–5	– Caregiver-child-detachment – Regulation of physiological processes, primary needs, take solid food – Language abilities – Motor skills: crawl, walk-control elimination of body wastes – development of basic autonomy – Describing social and physical reality – Forming concepts, playing	Playing, pretending, no logic thinking, but forming concepts, no manipulation of information mentally, egocentric stage	– Development in infancy seems to be inconspicuous – Growth deficit becomes obvious, use TS-specific growth charts Clinician should be aware of: – Specific cognitive phenotype and psychosocial phenotype in TS – Higher risk for ADHD, autistic traits If developmental deficits are suspected: – Screening for motor skills – Screening for neurodevelopmental deficits	Bailey scales, WIPPSI-III, NEPSY, KABC-II, SSIS, CBCL
Childhood 6–12	– Getting along with age mates – Physical skills necessary for ordinary games – Developing concepts necessary for everyday living – Developing conscience, morality and a sale of values – Masculine or feminine role – Developing attitudes toward social groups/institutions – Achieving personal independence	Ability to think logically, when concrete subjects involved, classification skills improve, egocentric view is over, viewpoints of others can be taken in account	– Ask for achievements in school – when difficulties are reported, organize specific support – Specific risk of impairment in visual memory, visual attention, visuospatial processing, organize specific support – Explore social behavior and social activities – when difficulties are reported: give recommendation for social skill training Girls with TS identify as female, puberty matters require diligent discussion and empathy – Start to explain normal pubertal development around age 10 years – Explain the need to induce puberty, if laboratory results point to ovarian failure around age 11 years	WISC, NEPSY, KABC-II, SSIS, CBCL, YSR, TRF
Adolescence 13–17	– Achieving a masculine or feminine social role – Accepting one's physique and using the body effectively – Achieving new and more mature relations with age mates of both sex – Achieving emotional independence of parents and other adults – Desiring and achieving socially responsible behavior	Thinking logically and abstract, able to use metacognition, problem solving in multiple steps develops	Psychosocial issues increase during adolescence, self-perception may be affected in a negative way: – Romantic love, sexuality and fertility matters should be addressed – Involve a specialized endocrine gynecologist for information on estrogen substitution, fertility matters Cognition: verbal skills are in normal range, but impairments in executive functioning, self-regulation, working memory, planning skills and abstract reasoning might emerge – Organize psychological testing and support (complex cognitive processes), if necessary Vocational planning should start – Support emancipating from parents, encourage health autonomy and self-care skills in an age-appropriate way (Lit Tabell zitieren)	WISC, WAIS, NEPSY, KABC-II, SSIS, CBCL, YSR, TRF, TRAQ

(Continued)

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Table 1 Continued.

Normative developmental concepts			Turner syndrome: pullouts for psychosocial care	Psychological tools
Age	Developmental tasks (Robert J Havighurst (20))	Cognition (Jean Piaget (21))		
Transition			– Follow the process of transition readiness, use questionnaires enquiring healthcare autonomy, self-care, disease management	TRAQ
Young adulthood 18+	– Selecting a mate, learning to live with a partner – Starting family, rearing children – Getting started in occupation – Taking on civic responsibility – Finding a congenial social group		Transition matters: concrete phase of starting to transition, contacting adult providers – Address sexuality and fertility matters Given social impairments in TS – Supporting young women and their families for living as independently as possible – Enhancing academic career, job training, professional life – Promoting hobbies, voluntary work, creative abilities, etc. can counteract social isolation and dependency on family	WAIS, KABC-II, SSIS, CBCL, YSR

parents and/or the patient about the various aspects of the diagnosis and treatment options (24). Experienced pediatric endocrinologists specialized in growth disorders should be involved in order to inform about the option of growth hormone (GH) therapy. The information provided should include the estimated benefit of approximately 7 cm in adult height (25) without overrating the effects on health-related quality of life (HRQL) (see below).

Accordingly, we advocate incorporating family counseling through a multidisciplinary care team immediately on disclosure of the diagnosis (7, 26). A structured and transparent documentation of the information disclosed and the family's reaction is a support for the pediatrician in enhancing the inevitable ongoing process of disclosure according to the age and developmental stage of the patient. At transition, starting at the age of 11–13 years, the clinician should repeat information about medical and health issues that were disclosed to the parents directly to the girl. Standardized documentation sheets are available for this purpose (27).

The parents might also need counseling support to counteract both the feeling of guilt, which has been reported in well-educated mothers of girls with TS (28), and the risk of poorer adjustment of parents reported in lower socio-economic classes (29).

Awareness of the psychological and psychosocial issues in TS is as important as the knowledge of treatment options. Importantly, information on the aspects of cognitive development should not instill fear of mental disability but stress the variability and strengths of cognitive development in TS (see below). Any developmental or neuropsychological screening procedures indicated should be initiated at an early stage and remain an integral part of the patient's routine treatment.

Furthermore, information about self-help groups, support groups and specialist literature may be helpful.

Infancy, 0–2 years

A divergence from normal psychosocial development is unobservable during infancy in girls with TS (30). However, height and weight typically drops below that of age-matched normal developing girls (31), often causing parental concern. Mothers especially worry about a child's failure to gain weight (32, 33, 34, 35) as weight gain is closely related to maternal responsibilities and abilities. Furthermore, even where there is no obvious aberration parents are anxious to receive advice on how best to nurture the child.

Recommendations

In the case of an antenatal or postpartum diagnosis of TS, a positive child-mother/father attachment is crucial and will be enhanced by diligent disclosure (see above). TS-specific growth charts (36) are useful for monitoring expected growth and weight gain and relieve mothers of guilt feelings.

Additionally, pediatricians should be attuned to sleeping and eating habits or emotional disturbances of the infant because irregularities stress the mother-child dyad and mothers might need support in adjusting to their baby's demands (37). Screening by using the Bayley scale (38) is strongly recommended as soon as delayed development is suspected.

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Early childhood, 2–6 years

The difference in height compared with age-mates becomes obvious in early childhood. However, there is no evidence of any difference in development between girls with TS and their age mates regarding basic achievements such as controlling body functions, playing and operating with simple cognitive concepts. Albeit, reduced attention spans and a preference for playing with younger children have been observed in girls with TS, which might be due to the specific cognitive phenotype in TS (described in detail in the paragraph childhood, age 6–12 years) that is more obvious at a later age and includes attention deficits, hyperactivity and poorer social competence (12, 39, 40).

Recommendations

If short stature of a girl with TS is reported to become an issue within the family, both the psychosocial and auxological aspects should be considered. The benefit of an early start of GH therapy for catch-up-growth aimed at the lower normal range or even normal range (16) should be weighed against the possible strain from daily injections. As there are no data about a benefit from an early start on final height, the authors are reluctant to start GH therapy before the age of 4 years in their clinic.

Given the evidence of a specific cognitive and psychosocial phenotype in TS, pediatricians should be attuned to upcoming developmental impairments and therefore explore the child's social behavior, playing habits, concentration levels, cognition and communication skills.

When peculiarities are reported, psychological counseling and/or screening by using the Bayley scale for development in general, Wechsler Preschool and Primary Scale of Intelligence (WIPPSI) (41) for cognitive development or A Developmental NEuroPSYchological Assessment (NEPSY-II) (42, 43) for assessing neuropsychological domains and the Social Skills Improvement System Rating Scale (SSIS) (44) respectively the Child Behavior Checklist (CBCL) (45, 46) for social behavioral assessment are recommended. Figures 1 and 2 give brief information on the psychological tests recommended.

Occupational therapy (47, 48) may be helpful when delays or disorders in the psychosocial development of a girl become apparent at this early age. Occupational therapists should give training to patients with TS to improve their sensory processing, promote functional and coping skills as well as advising and encouraging parents to adopt beneficial strategies.

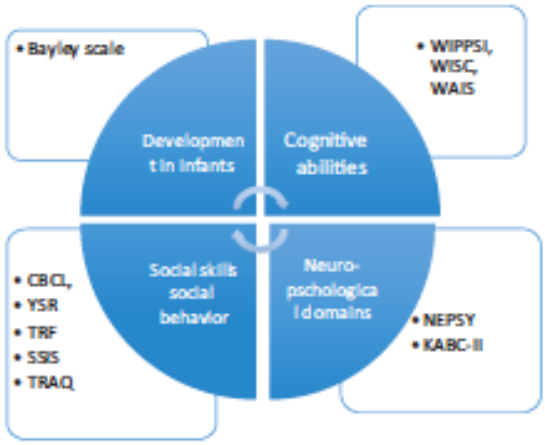

Figure 1
Psychological tests matched with areas of interest.

Childhood, 6–12 years

Physically, girls with TS grow slower and attain shorter height than their normally developing counterparts (4, 7), provoking comments from age mates and adults. Parents and children perceive the child's QoL, taking height into account, differently, parents rating it lower than the young girls do (49).

Studies describe TS-related impairment of motor abilities including fine motor skills, endurance in physical activities and body balance (50).

Most girls with TS show a normal Full Scale Intelligence Quotient (FSIQ) with no prevalence of mental or massive cognitive impairment. Mental retardation is observed only in approximately 10% of the girls, mostly those with a ring or marker chromosome (7). However, TS generally is associated with a specific cognitive phenotype with a consistent shift towards a higher Verbal Intelligence Quotient (VIQ) and a lower Performance Intelligence Quotient (PIQ) (11, 51). The girls' mathematical abilities are commonly impaired (14, 52). Furthermore, deficits in visuospatial processing, visual memory and visual attention become obvious between the ages of 6 and 12 years (12, 13, 53, 54, 55). Girls with TS are also at a higher risk for neurodevelopmental conditions such as attention-deficit hyperactivity disorder (56, 57, 58). These setbacks lead to problems at school (51). A total of 40–70% of the girls have been reported as having learning difficulties (7, 59).

Psychosocial problems also become obvious at this age. Poorer social skills have consistently been described as affecting all aspects of social behavior and functioning

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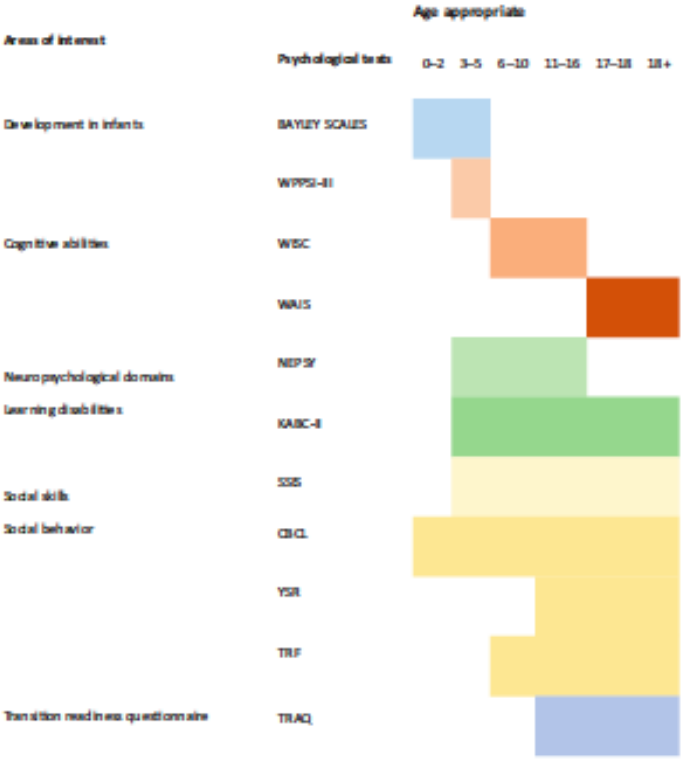

Figure 2
Overview of age-appropriate testing.

(13, 56, 60). The girls tend to have fewer close friends, spend less time with peers and appear emotionally less mature than age-matched normally developing girls. Building up friendships and maintaining relationships are difficult for them and parents report that their daughters with TS as less socially competent than their peers (13).

Recommendations

Reactions and comments from the social environment relating to the child's shorter height should be discussed between pediatricians and the parents as well as the child in an age-appropriate way to best cope with the child's increasing perception of being of a different height. This is the very latest point at which specialists in growth disorders should be involved to ensure parents receive state-of-the-art recommendations based on the latest findings in GH therapy also taking QoL outcomes into account.

Considering possible motor deficits also in adult age (61), we recommend encouraging girls with TS to undertake physical activities such as dancing, walking, swimming, hiking, biking, etc. and train their motor skills. Always bear in mind the special health condition of TS and ensure recommendations are in line with the girl's specific situation.

School progress is of major concern for parents and may greatly affect the girls' future QoL. Exploring school matters and recommending evaluation when difficulties with school subjects and/or concentration are reported is essential. Assessment using the WISC (62), the NEPSY (42, 43) or the Kaufman Assessment Battery for Children (KABC-II) (63) (Figs 1 and 2) is decisive for tailored cognitive remediation.

Furthermore, the way a girl interacts with peers provides information about age-appropriate social development. Screening for social behavior impairments by using the SSIS, CBCL or the Teacher Report Form (TRF) (45, 64) helps to explore these topics. Social skills intervention groups might help the patient if she has problems getting along with peers or is socially isolated.

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Adolescence, 13–18 years

Delayed puberty or the absence of pubertal development is the main medical and psychosocial issue during the phase of adolescent age and will lead to intensified medical care (3, 4). Additionally, during adolescence girls with TS increasingly fall behind in normal cognitive development. Tasks such as abstract reasoning, clustering and the use of metacognition are likely to become a challenge. Difficulties in mastering social cognitive tasks are especially evident when multiple cognitive domains are required (65). These findings correlate with deficits reported in other areas of cognition where executive functioning is involved (40, 66, 67). Limitations in working memory and slower response times indicate deficits in executive functioning that commonly result in weak planning, self-organization and self-regulation performances (12, 13, 39, 40, 53, 66, 68).

Verbal skills, however, are relatively strong in girls with TS, with normal to above average scores generally reported (11). Recent studies indicate a possible specific TS profile in language use abilities with increased vocabulary, initial verbal memory, reading comprehension and understanding of rarely used words. Sometimes reported slow responding and weak performances in verbal fluency tasks are less related to linguistic skills but rather reflect underlying deficits in executive functioning (69, 70, 71).

Adolescent girls with TS tend to have more difficulties than their age-matched peers to emancipate from their families, which might be due to increased anxiety, hyperactivity, impulsivity (72) or traits seen on the autism spectrum (55, 73, 74, 75). However, girls with TS differ in an important respect from patients with autism: they do desire social interaction (60).

Romantic love and sexual relationships are yet another sensitive topic emerging at this age (29, 56). Young girls with TS experience primary amenorrhea and impaired psychosexual development despite their female gender identification. Often, their first sexual experiences are delayed and subsequent experiences infrequent (76). Moreover, limited sexual experience is associated with lower self-esteem (29, 77).

Recommendations

The need for estrogen substitution to promote feminization is the central topic in the pediatric-endocrine setting at the end of the childhood stage or at the latest during early adolescence. Full information about the biological background of ovarian insufficiency in TS should be given again to the parents and in an age-appropriate manner to the girl.

Progress in education and vocational planning are decisive for the girls' future QoL as we will explicate below. Intensive exploration of cognitive and psychosocial development is essential. Reports of weak academic achievements and/or social problems are indications for psychological counseling or assessment. The psychological tests (WISC, NEPSY, KABC-II, SSIS, CBCL, YSR, TRF) we recommend are listed in Figs 1 and 2. Early intervention in school matters such as contacting and involving supportive teachers may be of great help.

We propose group training for social cognition and competence, where social skills are lacking.

Equally important are questions concerning romantic feelings, sexuality and fertility. As parents often have difficulties broaching these sensitive topics, healthcare professionals need to address them and provide in-depth information.

We recommend that special attention is given and extra resources are mustered to manage the sensitive phase of transition, see 'Health autonomy and transition' below.

Health autonomy and transition

Many difficulties and poor medical care of adult women with TS during the transition process have been reported (18, 78). Effective coordination and continuity of healthcare during a chronically ill adolescent's transition to adult care is critical for the patient's medical and emotional state in adulthood (17, 79, 80). The chances for a successful transition are decreased in some chronic conditions, including TS, because it is difficult to find experts in adult care (81). Young women with TS are furthermore seldom fully aware of their conditions (82, 83). In adult life, less than 4% of patients with TS undergo all the recommended medical investigations on a regular basis (84).

There are many reasons for transition failures such as poor self-advocacy or self-management, little family support or unsatisfactory cooperation between healthcare professionals and organizational structures (85). Self-care and healthcare autonomy, both considered preconditions for a successful transition, involve complex cognitive and regulatory dimensions (86, 87). Unfortunately, planning and self-organizational skills in girls with TS may be impaired even more than in other chronic conditions. Taking over responsibilities additionally to emancipation

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from their parents is likely to be a major hurdle for them. Therefore, extra effort and proactive action are required from healthcare professionals at this stage of development (81, 88).

Experts agree that a good QoL is the most important outcome. Building up a relationship of trust with new healthcare professionals, and having a social network are essential for a successful transition. Patients' understanding of their condition, its possible complications and medication as well as adherence to medication regimens and self-management are also seen as important individual contributors to achieve a good QoL (89).

Numerous transition programs have been introduced for a wide range of chronic diseases (90, 91, 92, 93, 94, 95) but, despite concerted efforts, results are still disappointing and transition in general is associated with poor health outcomes, non-adherence and no-shows at potential healthcare providers' appointments (96, 97).

Recommendations

Various transition models have been developed for chronically ill patients (98, 99). These commonly recommend informing the patients early (11–13 years) about health issues (27). Especially in complex diagnoses such as TS and where diagnostic disclosure may have taken place more than 10 years earlier, updating information on medical and health issues given to the parents and increasingly conversing with the patient alone is of crucial importance (27).

In addition, awareness of the process of 'transition readiness' has been recommended (100). In this process of transition, the developmental age is more important than the chronological age (4, 101), which is probably even truer for girls with TS, given their potential complex psychosocial impairments. Professionals should cooperate with parents in enabling adolescents to become more independent (102).

We recommend clinicians adopt the process of transition readiness. To our knowledge, no TS-specific transition readiness process has been developed. However, a disease-neutral questionnaire could serve this purpose. We recommend the Transition Readiness Assessment Questionnaire (TRAQ) (103) because of its economic features, clear structure and ease of use. It helps in deciding if the girl is ready for transition or whether education on health issues and an increase in healthcare autonomy is still needed.

Inclusion of a specialized gynecologist from the age of 16 years onwards is important to provide the women with information about current best practice in estrogen substitution and *in-vitro* fertilization. Once health autonomy has been gained and medical follow-up has been personalized, the young women will be dismissed from pediatric care. We recommend a tertiary center for the care after the patients' childhood and adolescence.

Adulthood

Adult women with TS not only face a decreased health status but also manifold psychosocial difficulties. Being unable to give birth and raise biological children causes grief in patients with TS. Although fertility aid is changing rapidly as research and possibilities in reproductive medicine develop (104, 105), still, the only slight chance of getting pregnant spontaneously and the health risk in pregnancy is of great concern for women with TS (106).

Height and the estimated benefit from GH therapy have been found to have no influence on QoL (107, 108, 109). Although overall QoL results have been normal in women with TS aged approximately 20–25 years, results of studies in women over 35 years of age are inconsistent (reviewed in (109)).

Women with TS show higher levels of alexithymia (110) and score lower in tests on self-perception and attitude towards their body than normally developing girls, even after they achieve a height in the lower normal range with growth hormone therapy (111). This negative self-perception might be the result of them being aware of atypical physical development in adolescence and perceiving the absence of spontaneous pubertal development (4).

A sexual relationship is known to help protect against feelings of loneliness or negative self-perception (28). Unfortunately, however, fewer women with TS than their age-matched peers report living in a sexual active partnership and in addition single women with TS report low sexual activities (112).

Adult women with TS tend to leave the family home later in life (113). More women with TS than age-matched control women remain unmarried and they report fewer social contacts (114). Lower psychosocial well-being seems to correlate with sex hormone deficiency but also with reported learning difficulties at school in the past (113).

The literature is contradictory on QoL associated with vocational training and employment. Less employment or

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employment below the women's vocational training lead to lower overall life satisfaction correlated to professional life than in control women (115). In contrast, some studies have reported educational achievements and job levels in women with TS equal (76) or even above that of the average female population in the United States (116).

Recommendations

Our proposed model of care during adulthood is for close collaboration between an endocrine gynecologist, cardiologist and internist with internal medicine becoming more and more important with increasing age.

Regarding family planning and raising children, information about various options should be given at the time of actual need and involve an informed team familiar with the options and risks of induced pregnancy after oozyte donation. Adoption should be considered as another way of fulfilling motherhood desires (117, 118).

Our task as healthcare providers must be to ensure young women with TS are well equipped to deal with the future challenges of their professional and social life. This includes instilling the importance of training for an appropriate career and considering integration into a social group outside their family of origin or romantic relationships. Importantly, the interests and particular qualities of women with TS such as creative abilities, enjoying voluntary work, activities close to nature, working with children, traveling etc. ought to be emphasized.

Psychological tests

As economic resources are usually limited, we have compiled a list of the most important physiological tests we would recommend that are also specifically reported in the literature on TS. These tests in our view identify cognitive, emotional and social deficits in a relatively short time with a low personnel cost.

Psychological tests we recommend:

- The Bayley Scales assess developmental levels of infants in the domains of motor skills, cognition, language, social-emotional and adaptive behavior (38).
- The Wechsler Preschool and Primary Scale of Intelligence (WIPPSI), the Wechsler Intelligence Scale for Children (WISC) and the Wechsler Adult Intelligence Scale (WAIS) assess verbal and non-verbal abilities (41, 119).
- The Developmental NEuroPSYchological Assessment (NEPSY-II) examines neuropsychological domains including social cognition and is useful in planning intervention for various childhood disorders (42).
- The Social Skills Improvement System Rating Scale (SSIS) locates social skills, competing behavior problems and academic competence (44).
- The Kaufman Assessment Battery for Children (KABC-II) identifies learning disabilities or developmental problems (63).
- The various forms of the Achenbach System of Empirically Based Assessment (45, 46) pinpoint problems such as depressive symptoms, anxiety or attention deficits in children and adolescents.
- The Child Behavior Checklist (CBCL) is a caregiver report form, whereas the Youth Self Report Form (YSR) is filled out by children and adolescents. The Teacher Report Form (TRF) may be of additional use for providing information in school matters.

More detailed recommendations on developmental tests are provided in the literature by Weitzman and coworkers (120) and the Council on Children with Disabilities (121).

Conclusion for the psychosocial care of girls with TS

Given the complexity of a TS diagnosis and the psychosocial impact of the condition, counseling on psychosocial issues and addressing concerns about the girls' daily life and future adult life needs to be integrated into standard pediatric endocrinologist care. Importantly, counseling should always be one step ahead. Accordingly, our recommendations mostly deal with potential upcoming or expected difficulties in growing up with TS. They aim to enhance pediatricians' preparedness while incorporating support from psychologists or other healthcare providers for optimal treatment of girls with TS.

Centers that treat girls and young women with TS are challenged to develop a supportive mode of transition to increase the chances for patients with TS to receive the best medical care and psychosocial support throughout their adulthood. Health autonomy is of central importance for women with TS as tertiary centers for rare diseases are imminently to be established for diagnosis made during childhood.

We contend that a multidisciplinary standard of care based on well-defined screening would greatly improve

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the long-term outcomes of patients with TS. Until such a standard is universally established, we are convinced that if account is taken of the considerations we have set out; even practitioners acting on a lower economic level will be able to improve the long-term outcomes for patients with TS.

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The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of this review.

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2.2. Be on TRAQ — Cross-cultural adaptation of the Transition Readiness Assessment Questionnaire (TRAQ 5.0) and pilot-testing of the German Version (TRAQ-GV-15)

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Transition from pediatric medical settings to adult-centered medical care has been gaining growing importance within the last years. Health outcomes of children and adolescents with chronic conditions are improving. Survival rates are increasing due to medical advance. Therefore, the change of medical care, the transfer to adult care, is of utmost importance. Adolescents and their families are confronted with the process of transition to a new health care setting. However, many young patients are either lost for follow-up or show a low level of medication therapy treatment adherence in young adulthood. Until recently, adolescents were released from pediatric care at the age of eighteen. More recent literature recommends assessment of readiness for transition using validated questionnaires. No validated transition readiness assessment questionnaire was available in German language. Therefore, I decided to cross-culturally adapt an internationally recognized assessment instrument, the TRAQ 5.0 and to pilot-test the German version TRAQ_DV_cc_Version 2.1. April 2017 for psychometric properties. This work resulted in the TRAQ-GV-15, a clinically valuable and easily applicable screening tool for use in the course of transitioning adolescents in the need of medical follow-up in adulthood.

My contribution to this project was given in the conception and the designing of the study, in building up the cooperation with the American authors and the collaboration with the colleagues in Freiburg, Germany. I contributed in writing applications for ethical approval and grant applications, monitored the translation and discussed the drafts with our American partners. Further, I was involved in collecting data, I conducted statistical analyses and drafted the manuscript.

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Original Article

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Be on TRAQ – Cross-cultural adaptation of the Transition Readiness Assessment Questionnaire (TRAQ 5.0) and pilot testing of the German Version (TRAQ-GV-15)

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Abstract: Transfer from pediatric care into the adult health care system is known to be a vulnerable phase in the lives of youth with special health care needs (YSHCN). Recommendations from the literature favor assessment of transition readiness rather than simply pass over YSHCN from pediatric to adult-centered care by the age of 18. Nevertheless, no validated and disease neutral assessment instrument exists to date in German. Hence, our aim was to cross-culturally adapt and to pilot-test a German version of the Transition Readiness Assessment Questionnaire (TRAQ 5.0). We wanted to provide a tool that can be applied broadly during the health care transition (HCT) process of YSHCN. The development included translating and adapting TRAQ 5.0 to German and conducting a pilot-study with 172 YSHCN between the ages of 14 and 23. Cross-cultural adaptation resulted in the TRAQ-GV-15. Exploratory factor analysis led to a 3 factor-structure. Internal consistency for the overall score was good with a Cronbach's alpha of 0.82. Age, in contrast to sex, had a significant effect on the TRAQ scoring. The administration of the TRAQ-GV-15 was well received and demonstrated good feasibility.
Conclusion: The TRAQ-GV-15 is an easily applicable and clinically usable instrument for assessing transition readiness in German speaking YSHCN prior to HCT.

Keywords: autonomy; health care self-management; health care transition; health literacy; pediatric onset chronic illness; youth.

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Introduction

The transfer from pediatric care to adult care carries a considerable health risk for youth with special health care needs (YSHCN [1–3]. This phase is associated with missed medical appointments [4, 5], poor health outcomes [6] and lower overall quality of life [7, 8]. Recommendations include structural changes in health care systems [9–11] as well as propositions on how to support patients while transferring [12–17]. Although various transition programs have been introduced [18–21], no gold standard in transitioning YSHCN from pediatric care to the adult centered care has been established so far [11, 22–24]. Nevertheless, an urgent need for enhanced success in transitioning is evident in the field of chronic and rare diseases [25, 26]. Internationally, experts are working on the implementation of standardized transition processes at pediatric clinics [27, 28].

Transfer is usually scheduled at the age of 18 [29–31]. In some practices, youth remain in pediatric care until the age of 24. Recent literature emphasizes focusing on readiness for transition [32] rather than on chronological

age [33] and recommends to regularly assess and evaluate disease self-management and health literacy skills in YSHCN by objective and validated tools [7, 34]. However, psychometrically sound and reliable instruments are sparse [35, 36].

In follow-up of these recommendations, we performed a literature search, no validated disease neutral transition readiness assessment instrument was to be found in the German language. However, we encountered an instrument in the English language, developed in the USA – the TRAQ 5.0 (Transition Readiness Assessment Questionnaire) [37, 38]. The TRAQ has demonstrated good reliability and validity [34], and we considered this tool as adequate for our actual needs. Meanwhile, initial intervention trials using the English TRAQ have been conducted [39, 40]. Furthermore, several other cross-cultural adaptations of the TRAQ have been carried out recently [41–43]. This study is the first adaptation of the TRAQ 5.0 for German speaking YSHCN.

Material and methods

Description of the original instrument

The TRAQ 5.0 [38] is a disease-neutral, self-administered questionnaire assessing transition readiness in adolescents and young adults (AYA) aged 12–26 years with chronic conditions and therefore with special health care needs. It comprises 20 items subdivided in 5 subscales ("Managing Medication", "Appointment Keeping", "Tracking Health Issues", "Talking With Providers" and "Managing Daily Activities"). The response set of the English TRAQ is derived from the Transtheoretical Model [44]. Nevertheless, the items score in a range from 1 ("No, I do not know how") to 5 ("Yes, I always do this when I need to") with a maximum total score of 100 and a minimum total score of 20. A lower overall score indicates less transition readiness.

Translation and cross-cultural adaptation of the TRAQ

After approval of the authors of the TRAQ 5.0, the cross-cultural adaptation of the questionnaire was conducted according to recommendations from the pertinent literature [45]. Initially, two bilingual health care professionals from the two study sites (Vienna, Austria and Freiburg, Germany) familiar with the TRAQ 5.0 independently translated the original 20-item TRAQ 5.0 to German. Then, two English native speakers (US American and Canadian, fluent in German) both blinded to the TRAQ and not otherwise involved in the study, independently translated it back to English. In a third step, we discussed our back-translated English versions with the authors of the TRAQ 5.0 via skype. Equivalents and discrepancies due to different health systems needed some considerations. Areas that lacked consistency or clarity were discussed. We looked for ways to translate English phrases and colloquialisms in German. These edits were incorporated into the German version. In the original American TRAQ 5.0 five items (item 14 and item 15 as well as item 18, item 19 and item 20, all three items assigned to the "managing daily activities" scale) did not add statistically relevant information in interpreting the TRAQ results: item 14 and item 15 showed low factor loadings, and furthermore, the "daily activities" subscale showed a lower Cronbach's alpha than the other subscales. Therefore, the 5 items were omitted in the German Version of the TRAQ.

Hence, we obtained an instrument with 15 items where scores fall between maximum 75 and minimum 15. Finally, we pilot-tested the questionnaire for comprehensibility in a convenience sample of ten YSHCN aged 14–19 at the outpatient clinic of the Medical University of Vienna. Minimal changes led to our definite version, TRAQ-GV-15.

Survey sample

Our study population comprised AYA under medical care at two outpatient clinics, the Department of Pediatrics and Adolescent Medicine, Medical University of Vienna and the Center for Pediatrics, Medical Centre – University of Freiburg, Germany. Both clinics provide comprehensive primary care services for a broad variety of chronic conditions. The University of Freiburg is linked to a nationwide transition program [46], but no standardized transition readiness assessment has been implemented so far. In both clinics care providers can extend medical treatment of patients beyond the age of 18 if they think that a transfer to adult care in the present state would pose a health risk to the patients. Hence, the resulting convenience sample from the two study sites consisted of 194 YSHCN aged from minimum 14 to maximum 23 years of following outpatient clinics: Gastroenterology, Nephrology, Endocrinology (Diabetes, Growth and Bones), and Pulmonology. Exclusion criteria were severe cognitive impairment or poor German language skills.

Questionnaire administration

The questionnaire was administered to YSHCN at the two study sites from March 2017 to October 2017. The responses

to the self-administered paper-and-pencil survey were collected during routine clinical care. Participants were instructed to answer the items on the survey on their own. However, a study investigator assisted whenever support was needed. Clinical data were obtained from medical records. Demographic data such as socioeconomic and family background, first language and educational level of the patients, were obtained by means of an additional anamnestic questionnaire. Supplementary information related to the feasibility of the TRAQ like "help needed", "fill-in time" and "consulting time" were documented by the study investigators.

Statistics

We tested the TRAQ-GV-15 for reliability, construct validity, and scale-internal consistency. We applied the class mean imputation method for corrected mean sums to compensate for missing data. However, less than 0.5% of the values of any given item were missing. We conducted standard descriptive statistics for single items and overall TRAQ-GV-15 score description including frequency distribution, means, standard deviation, minimum, maximum and median [47]. To test validity for age and sex we conducted an ANOVA with Bonferroni adjustment for multiple comparisons [48]. A p-value of <0.01 was considered as statistically significant.

Because we eliminated five items from the original TRAQ 5.0 there was no evidence for a beforehand structure of our adapted 15 items instrument. Therefore, we conducted an exploratory factor analysis (EFA). We verified adequacy of the sampling with Kaiser-Meyer-Olkin- and Bartlett's-Test. We conducted Principal Component Analysis and Orthogonal Varimax Rotation with Kaiser Normalization. Internal consistency of the German questionnaire was estimated using Cronbach's alpha [47]. Statistical analysis was performed with IBM® SPSS® software Version 22.

Results

Sociodemographic characteristics of study sample

The final sample comprised the data sets of participants with given informed consent (n = 172). We included patients with following conditions: Cystic Fibrosis (n = 24), Type 1 Diabetes Mellitus (n = 76), Juvenile Idiopathic Arthritis (n = 49), Chronic Renal Diseases (n = 12) and various rare conditions such as Turner syndrome or Glycogen Storage Disease (n = 11). Mean age of our population was 16.9 years (SD ±1.8) ranging from 14 to 23 with almost two thirds female respondents (60.5%). More than half of our respondents (62.2%) reported that German language was their first or main language, all participants however were fluent in German. Information on insurance status was not included in our survey since general social insurance applies to all our patients. Data on educational level and school type were either lacking (26.7%), inaccurate (6.4%) or grouped as middle school level (8.7%), as vocational training (7.6%) or as high school level (50.6%). Sociodemographic characteristics are listed in Table 1.

Feasibility

Feasibility was tested by assessing if whether or not respondents could answer the questionnaire without help, and whether or not the questionnaire was completed within 10 min or if participants needed extra explanation in addition to a brief introduction. The majority (76.7%) completed the TRAQ-GV-15 within 5 min, about 20% within 10 min, respectively. Only 24 respondents (13.9%) needed help more than once. It became apparent that 15 min of explanation and introduction were sufficient for most participants (78.5%). Data shown in Table 2.

Table 1: Sociodemographic characteristics of the study sample included in the validation of the German version of the Transition Readiness Assessment Questionnaire (TRAQ-GV-15).

n	172	100.0%
Age		
14–15.99	55	32.0%
16–17.99	68	39.5%
18–23.99	49	28.5%
Sex		
Female	104	60.5%
Male	68	39.5%
Diagnosis		
CF	24	14.0%
DM	76	44.2%
JIA	49	28.5%
NE	12	6.9%
RD	11	6.4%
Language		
German	107	62.2%
Eastern Europe	28	16.3%
Western Europe	3	1.7%
Asia	10	5.8%
No data	24	14.0%
Type of education		
Middle school	15	8.7%
Vocational training	13	7.6%
Highschool	87	50.6%
Not specified	11	6.4%
No data	46	26.7%

Table 2: Feasibility of the administration of the TRAQ-GV-15.

Aspects assessed	n = 172	%
Help needed		
0–1 times	148	86.1%
2–3 times	20	11.6%
4+ times	4	2.3%
Time for completion		
2–5 min	132	76.7%
6–10 min	34	19.8%
10–20 min	5	2.9%
More than 20 min	1	0.6%
Time for counselling		
None	6	3.5%
Up to 15 min	135	78.5%
Up to 30 min	28	16.2%
Up to 60 min	2	1.2%
More than 60 min	1	0.6%

TRAQ-GV-15, Transition Readiness Assessment Questionnaire, German version, 15 items.

Exploratory factor analysis

EFA was conducted on our sample of 172 respondents to assess a factor structure of the TRAQ-GV-15. Model adequacy was verified with Kaiser-Meyer-Olkin (0.823) and Bartlett's-Test ($p < 0.001$), indicating suitability for factor analysis. In the first round of EFA (data not shown) using Orthogonal Varimax Rotation, four factors demonstrated an Eigenvalue ≥1 explaining 54.94% of the total variance. The scree plot showed a clear elbow with only the first factor, demonstrating an Eigenvalue of 4.38. Furthermore, factor 4 comprised only two items. Analysis of reliability is not meaningful with solely two items [47]. Therefore, we conducted another round of EFA extracting three factors accounting for 47.87% of the total variance. The new factor structure led to three clinically meaningful domains for the TRAQ-GV-15: domain 1 "Autonomy" (items 01, 04, 05, 07, 12, 13, 15), domain 2 "Health Literacy" (items 02, 09, 10, 11) and domain 3 "Adherence" (items 03, 06, 08, 14,). All items loaded on at least one factor at a level of more than 0.40 except item15 ("Do you answer questions that are asked by the doctor, nurse or clinic staff", factor loading 0.388). Factor loadings are demonstrated in Table 3.

Internal consistency

We calculated Cronbach's alpha Coefficient for internal consistency. The TRAQ-GV-15 presented globally a Cronbach's alpha = 0.824 which indicates a good reliability [47]. For the three subscale domains, the values of Cronbach's alpha appeared as follows: Domain 1 Cronbach's alpha = 0.779, Domain 2 Cronbach's alpha = 0.721 and Domain 3 Cronbach's alpha = 0.507. Results are presented in Table 3.

Table 3: Cronbach's alpha and factor loadings of the TRAQ-GV-15 overall Cronbach's alpha 0.824.

	Traq domain 1 autonomy	Traq domain 2 health literacy	Traq domain 3 adherence
n	166	170	169
Cronbach's alpha:	0.779	0.721	0.507
Factor loadings			
Item05	0.822		
Item01	0.692		
Item04	0.654		
Item07	0.622		
Item12	0.596		
Item13	0.422		
Item 15	0.388		
Item10		0.776	
Item09		0.774	
Item11		0.759	
Item02		0.489	
Item14			0.671
Item08			0.587
Item06			0.568
Item03			0.485

TRAQ-GV-15, Transition Readiness Assessment Questionnaire, German version, 15 items.

TRAQ-GV item values and overall score

In line with the original TRAQ, the items of the TRAQ-GV-15 score in a range from 1 to 5. Of the expected 2580 (172 responses to 15 items) values only 12 values were missing which corresponds to 0.47% missing values. We calculated the TRAQ-GV-15 mean overall item score (54.77 ± 10.17) with SD (Table 4). Similarly, we analyzed the 15 items individually, see Table 5. Item11 ("Do you manage your money and budget household expenses – for example: use checking/debit card?") demonstrated the lowest mean

Table 4: TRAQ-GV-15 overall mean score for age groups and sex.

	Mean	SD
TRAQ overall	54.77	10.17
Age group		
14.0–15.99	48.52**	9.48
16.0–17.99	55.86	7.78
18.0–23.99	60.27	10.29
Sex		
Female	55.44	10.00
Male	53.76	10.43

*p-value of < 0.01; **p-value of < 0.001.

Table 5: Descriptive statistics of the 15 TRAQ-GV items.

Item	n	msc	SD	min	max	median
Item01	170	3.22	1.45	1	5	4
Item02	171	3.63	1.50	1	5	4
Item03	170	4.75	0.56	2	5	5
Item04	171	3.61	1.42	1	5	4
Item05	169	3.52	1.36	1	5	4
Item06	171	3.85	1.33	1	5	4
Item07	171	3.90	1.35	1	5	4
Item08	172	3.95	1.32	1	5	4
Item09	171	2.32	2.09	1	5	2
Item10	172	2.36	1.52	1	5	2
Item11	172	2.28	1.50	1	5	2
Item12	172	4.53	0.93	1	5	5
Item13	172	3.94	1.36	1	5	4
Item14	172	4.35	1.02	1	5	5
Item15	172	4.66	0.62	1	5	5

msc, mean scores; SD, standard deviation; min, minimum; max, maximum.

score (2.28 ± 1.50) whereas item03 ("Do you take medications correctly and on your own?") demonstrated the highest mean score (4.75 ± 0.56).

The data followed a normal distribution, no skew was found. The two-way (2 × 3) ANOVA showed no significant difference for sex. However, age had a significant ($p < 0.001$ after Bonferroni adjustment for multiple comparisons) effect on the overall TRAQ score. Adolescents of the youngest age group (14–15.99 years) scored significantly lower than YSHCN of the two other age groups comprising adolescents and young adults from 16.00 to 17.99 and 18.00–23.99 years. TRAQ overall mean scores were 48.52 vs. 55.86 and 60.27, respectively (Table 4).

Discussion

This work followed the urgent need [5, 10, 25] of improving the preparation for transfer of YSHCN to the adult medical care. Objective of the study was to translate and to adapt the English TRAQ 5.0 [38] for use in German speaking countries. Thus, the process of adaptation resulted in the TRAQ-GV-15.

For cross-cultural adaptation and pilot-testing of the TRAQ-GV-15, we collected data from 172 YSHCN at two academic pediatric clinics in Austria and Germany, respectively. Administration proved to be easy and feasible. Most of our respondents finished the survey in less than 10 min. The low number of missing values may be the result of face-to-face intervention including the offer of support whenever needed. We also collected data on parental education, family status of the parents and family income. Unfortunately, this data were incomplete or contradictory. Therefore, we could not include them into our analysis.

EFA resulted in 3 subscales. In our opinion, the newly defined 3 subscales (Domain 1: "Autonomy", Domain 2: "Health Literacy" and Domain 3 "Adherence") reflect domains contributing to transition readiness [37, 38].

Our TRAQ-GV-15 differs from the TRAQ 5.0 in some aspect. First, the TRAQ 5.0 study was a re-evaluation of the 29-item TRAQ whereas our study was the first adaptation in German language of the TRAQ 5.0 and a pilot-study of the TRAQ-GV-15. Next, the TRAQ 5.0 includes 20 items, while the TRAQ-GV-15 comprises 15 items. Moreover the English TRAQ has a 5-subscale structure, as opposed to the 3-subscale structure of the German version. Concerning psychometric properties, internal consistency of the TRAQ-GV-15 was lower for both the overall score (0.94 vs. 0.82) and for the subscales compared to the original TRAQ 5.0. Subscale 1 and subscale 2 of the TRAQ-GV-15 subscales indicated an acceptable internal consistency (0.78 and 0.72) whereas the third subscale demonstrated poor internal consistency with a Cronbach's alpha coefficient under 6.0. Finally, comparing criterion validity, we did not find significant effects of sex on the TRAQ-GV-15 mean scores after adjusting for age. This contradicts findings in earlier TRAQ studies [37, 38, 41]. In line with previous studies [35, 37, 38, 41], higher age increased TRAQ-GV-15 scores in our sample.

This finding underpins that transition readiness improves with age [38]. Nevertheless, individual maturation follows an unequal pace and needs positive reinforcement especially in YSHCN as health risk behaviors are known to be linked to chronic conditions [49]. Thus, assessing transition readiness in YSHCN could provide a good basis to discriminate between YSHCN ready for transition and AYA in need of intensified attention. Literature indicates that attitudes such as compliance and adherence are strongly linked to individual support [50].

Consequently, clinical value of our study results from raising awareness for the process of HCT in providers, patients, and parents. Furthermore, the TRAQ-GV-15 could be used to identify target areas for patient education and to enhance self-reliance in managing the disease. The repeated use of the TRAQ-GV-15 could help to evaluate the individual transition readiness process of adolescent patients.

Future studies will have to further evaluate the psychometric properties of the TRAQ-GV-15 among larger study samples. We realize that subscale three needs additional study. Of utmost interest is the predictive validity of the TRAQ-GV-15, which will have to be assessed in follow-up and longitudinal studies measuring transition

outcome. Eventually, upcoming studies will give clear evidence of what scores to expect from youth differing in age, sex and condition. Longitudinal data might widen our understanding of the development of transition readiness and could eventually lead to the establishment of cut-off scores, helping to identify patient samples at risk for potentially failed HCT timely [1]. The effect of intervention programs on TRAQ scores might also be worth investigating. Goal setting embedded within tailored interventions is another way to evaluate or improve TRAQ outcomes.

Limitations

The TRAQ-GV-15 is a self-reporting questionnaire which always holds the risk of socially-desirable answers. Moreover, the possibility of underestimation or rather overestimation of one's own competencies and skills is a frequent phenomenon. External observation like expert-rating or caregiver's assessment can compensate for these limitations. However, we did not include an external validation in our study design because we aimed at cross-cultural adaptation and validation of the TRAQ-GV structure.

Non-response was primarily caused by the fact that parents did not give informed consent. Therefore, we could not include data collected from YSHCN (n = 20) who managed their hospital appointments without parents or otherwise caregivers. Consequently, we had to exclude AYA who were ready to meet their doctor's on their own before the age of 18. This might attribute to a bias in our data.

Conclusions

We believe that our German version of the TRAQ has a direct benefit for YSHCN. In our experience, the TRAQ-GV-15's administration inevitably led to transition centered communication with health professionals, encouraged caregivers to enhance AYA's autonomy and sensitized YSHCN for transition specific issues. Based on the results of our study, we are convinced of the viability of the implementation into daily hospital routine.

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Author Contributions

Caroline Culen contributed to the conception and design of the study, to writing grant applications, to data collection and assessment, to statistical data analysis, interpretation of results, drafted and revised the manuscript, approved the final version of the article submitted. Marion Herle contributed to study conception, to managing and conducting the study, to interpreting the findings, critically revised the manuscript and approved the final version of the article submitted. Marianne König contributed to the translation and cultural adaptation of the TRAQ, contributed to conducting the study and gave final approval of the version submitted. Sophie-Helene Hemberger und Sanja Seferagic contributed to conducting the study and to data collection, critical revision of the manuscript and gave final approval of the version submitted. Carolin Talaska contributed to the translation and cultural adaptation of the TRAQ, to data collection and gave final approval of the version submitted. Diana-Alexandra Ertl contributed to conducting the study, final approval of the version submitted. Gudrun Wagner critically revised the manuscript and approved the final version of the article submitted. Christine Straub contributed to the translation and cultural adaptation of the TRAQ, to data collection, critical revision of the manuscript and gave final approval of the version submitted. Kiana Johnson and David L. Wood contributed to conception and design of the study, supplied to the translation and cultural adaptation of the TRAQ, critical revision of the manuscript and gave final approval of the version submitted. Gabriele Häusler was the PI of the study, contributed to planning and designing the study, to writing grant applications, supervised the study and had final responsibility, approved of the final version of the manuscript submitted.

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Informed consent: Both, parental consent and informed assent were mandatory for all participants under the age of 18. Participants older than 18 years gave informed consent. No parental consent was required.

Ethical approval: The study was approved by the ethical committee of the Medical University of Vienna (EK Nr. 1456/2016) as well as the institutional review board of the Medical Centre – University of Freiburg, Germany (369/17).

Participation was voluntary, adjusted study information was handed out.

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2.3. Less ready for adulthood? – Turner syndrome has an impact on transition readiness

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Research paper in preparation

Young women with TS are especially at risk to be lost for follow up after being released from pediatric care. Literature indicates that transition holds unequal challenges gfor different chronic conditions. Patients with more common chronic conditions without cognitive and psychosocial impediments coming along with the primary diagnosis seem to better master the transitional phase than conditions with neurocognitive challenges. Assessment of transition readiness has been recommended by researchers in order to enhance the preparation of the transfer from pediatric to adult medical care. Therefore, we conducted a study to investigate possible differences in transition readiness measured according to the TRAQ-GV-15 between TS patients and other chronically ill patients. TRAQ-GV-15 scores of twenty-seven girls and young women with TS were compared to the scores of twenty-seven age-matched girls with type 1 diabetes or a childhood onset rheumatic condition. The results show lower transition readiness scores for AYA with TS. Moreover, TS patients needed significantly more time to complete the questionnaire, which underlines the need for more time or personal resources in treating patients with TS.

My contribution to the project comprised writing applications for ethical approval and grant application, collecting and processing data and statistical analyses. I contributed in drafting and revising the manuscript.

Less ready for adulthood? – Turner syndrome has an impact on transition readiness

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Abbreviations:

AYA — Adolescents and Young Adults
HCT – Health Care Transition
TS — Turner syndrome
SD — standard deviation
YSHCN — Youth with Special Health Care Needs

Introduction

Transition paths are not yet clearly marked for patients with TS (1,2) and transition outcomes proved to be poor in the past (3,4). Appropriate medical care after transition is only found in 3.5% percent of TS patients (5,6). As the process of transfer from comprehensive pediatric to adult medical care has been studied more closely within the last years, considerations and recommendations for successful transition are becoming theoretically underpinned (7,8). Assessment of readiness for transition in the preparatory process has been recognized as beneficial for the imminent transfer of AYA to adult medical care (9,10). At present, assessment of readiness for transition with disease specific and non-specific questionnaires evolves to a more common practice internationally (10–12). However, recent literature points out, that the type of disorder might account for differences in transition readiness levels (13). Complex chronic conditions with neurocognitive involvement and delayed developmental maturation affect successful transfer to adult care for YSHCN (14). TS is known to be a condition with mild neurocognitive challenges and difficulties in social behavior (15). In addition, puberty is often delayed and personal maturity sets on later in life in young girls and women with TS. This is associated with decreased self-confidence and with a lower degree of autonomy than peers (16).

The main objective of the study was to examine whether individuals with TS show differences in transition readiness scoring compared to an age- and gender-matched population diagnosed with chronic conditions, such as(type 1 diabetes or childhood onset rheumatic disease (17,18). We hypothesized that girls and young women with TS would have lower indicators for transition readiness than AYA with type 1 diabetes or a juvenile rheumatic disease, namely when it comes to skills that include self-management, self-organization and self-advocacy.

Patients and methods

Patients

Our study sample comprised 54 adolescent patients treated at three specialized pediatric endocrine outpatient clinics. All three clinics provide comprehensive care for

patients with Turner syndrome according to international (19) guidelines and also to a broad range of childhood onset chronic conditions including DM Typ1 and childhood onset rheumatic conditions.

Inclusion criteria were adolescent girls and young women born between 1994 and 2003 and diagnosed with either TS, type 1 diabetes or with a rheumatic disease, more than six months prior to the start of the study. Exclusion criteria were lack of German language skills and/or severe cognitive impairment.

AYA were recruited from the registry of the pediatric departments. Patients who seemed eligible to the study were contacted via email/ telephone or were asked personally during routine clinical visits. After full explanation of the purpose and the course of the study, girls under the age of 18 gave informed assent, their parents provided informed consent. From young woman aged 18 to 23 informed consent was obtained.

The final study group comprised 27 female patients with TS. 27 age- and gender-matched participants diagnosed with type 1 diabetes or with a rheumatic condition (control group).

Clinical data for the TS group, such as treatment with growth hormone during childhood and adolescence, pubertal development and oestrogen substitution therapy, age at diagnosis, karyotype were collected from the patients charts.

Methods

Study design

Patients were either seen during routine clinical visits or invited for extra appointments at the study sites in Vienna or Graz.

Thirty-two patients with TS matching the inclusion criteria were identified at the three study sites. Contact was established in all cases, 28 patients responded positively. All 28 TS patients took part in the study. One girl had to be excluded because parent´s consent was obtained too late.

Thirty-eight female patients with type 1 diabetes and forty-three female patients with a rheumatic disease were identified. Contact was established in all cases. All individuals responded positively and took part in the administration of the TRAQ-GV-15. In our

study, comparison of TRAQ scores was conducted in two steps: first, we compared a group of female patients diagnosed with type 1 diabetes (n=29) to an age-matched group of female patients with a rheumatic condition (n=29) Comparison between the two groups showed no differences regarding transition readiness scores measured by the TRAQ-GV-15 and handling of the questionnaire (data not shown). This patient cohort served as control group for the TS patients. Second, we age-matched the TS participants (n=27) with the control group (n=27) and compared the data regarding transition readiness scores and application of the questionnaire.

Information on the individual handling of the TRAQ instrument was gained by supplementary documentation on "*fill-in time",* "*help needed*" and "*consultation time*" by the investigators. Data on first language, educational level and number of hospital visits per year were gathered by a sociodemographic questionnaire.
The study was conducted between March 2017 and August 2018.

TRAQ-GV-15

The German version of the TRAQ 5.0 (9) comprises 15 items within three subscales: autonomy (subscale 1), health literacy (subscale 2) and adherence (subscale 3). TRAQ-GV-15 item scores range on a 5-point Likert scale between 1 to 5, total TRAQ-scores fall between maximum 75 and minimum 15. Lower scores indicate less readiness for transition.

Statistics

Power and sample size analysis was performed (20). Differences between overall TRAQ scores, subscale scores and single item scores of the TRAQ-GV-15 were analyzed by non-parametric Wilcoxon signed rank test, in case of non-normal distribution. We used the Mann-Whitney U Test to test for significant differences between the two patient samples in handling the TRAQ questionnaire regarding fill-in time, frequency of help needed and consultation time (21). We considered a p-value of < 0.05 on a two-sided level as statistically significant. Analysis was performed with IBM® SPSS® software Version 22.

Results

TS study group: Characteristics

Participants characteristics are shown in Table 1. Average age at the time of the testing was 17.43 (*SD*±2.28) years in the TS group and 17.60 (*SD*±2.60) years in the control group. 4/27 were not treated with growth hormone , 4/27 had spontaneous puberty. One participant was diagnosed in her origin country and detailed information on history of estrogen substitution or spontaneous puberty was not obtained. The rest of the participants underwent pubertal inducement and estrogen substitution. The average age at pubertal inducement was 13,1 years old. All participants in the control group except one were presenting with a normal pubertal development for age and required no hormonal substitution.

Transition readiness indicators

We compared total TRAQ-GV-15 scores and scores within the three TRAQ-GV-15 subscales with SD, means, minimum and maximum between TS patients and controls. Computing the non-parametric Wilcoxon signed rank test, these results demonstrated a significant difference (p = 0.02) between the two groups, with lower overall scores for AYA with TS condition. In the TS group, mean overall TRAQ-score was 3.65 (±0.64), in the control group 3.95 (±0.68) (see Table 4).

Analysis of the three subscales indicated differences on a significant level (p = 0.009) for subscale 1 “Autonomy“ (TS group 3.81±0.88 vs control group 4.23±0.77). Differences for subscale 2 “Health Literacy” (TS 2.47±1.01 vs controls 2.91±1.28) and subscale 3 “Adherence” (TS 4.52±0.59 vs controls 4.53±0.64) showed no significantly different results between the two groups (Table 2).

Comparison of the two groups on a single item level displayed significant differences for the scores of three items: Item 05 (“Do you call the doctor’s office to make an appointment?”) and item 07 (“Do you arrange for your ride to medical appointments?”), both belonging to subscale 1 “Autonomy”, presented lower scores for TS AYA with a moderate effect size of r=.40 (Item05: TS 3.04±1.48 *mdn* 3 vs. controls 3.96±1.32 *mdn* 4 and item07: TS 3.19±1.55 *mdn* 3 vs. controls 4.15±1.32 *mdn* 5). Item 09 (“Do you apply for health insurance if you lose your current coverage?”) belongs to subscale 2

"Health Literacy". Scores were significantly lower for TS patients with a small effect size of *r*=.29 (TS 1.85±1.2 *mdn* 1 vs. controls 2.67±1.64 *mdn* 2) (Table 3).

Handling of the TRAQ-GV-15

Statistically significant differences could be found within "fill-in time" and "time of consultation". Less than 55% of the TS group finished the questionnaire within 5 minutes. A third of the TS patients answered the survey within ten minutes, but nearly 17% needed up to twenty minutes for the task. In contrast, 85% of the controls completed the TRAQ-GV-15 within five minutes, 15% needed up to ten minutes for filling out.

In regard of the consultation time, for 8% of the TS patients less than fifteen minutes of consultation time were documented, whereas 60% of the TS group needed thirty minutes of counseling and for 26% of the TS participants sixty minutes of counseling were documented. For controls, 85% of the AYA consumed around fifteen minutes, four youth up to 30 minutes. No significant difference between the two groups was found with respect to "*help needed*" in filling out the TRAQ (Table 4).

Analyzing the supplementary survey for patient characteristics, we could not test for significant differences due to lack of statistically adequate data. However, we report observed differences between the two groups in percentage. One or two clinical appointments per year were stated from 72.7% of the TS patients and from 10.5% of the control group. Three or four doctor visits per anno were reported from 27.3% of the participants in the TS group, whereas 89.5% o the controls reported three, four or even more routine clinical visits per year (table 5).

When assessing for the "first language" of our cohorts, 83,3% in the TS patient group and 66,6% in the control group reported German as their first language. Other first languages recorded were Turkish, French, Serbo-Croatian or Arabic (Table 5).

In the TS group 58,3% of the girls documented attending a secondary school, respectively high school. One girl was in middle school, two girls were in apprenticeship and six young women (25%) were having a job or stated further educational trainings (eg. Drama school, university). In the control group 84,2% of the girls went to high school, two girls were in apprenticeship. No other form of education was reported (Table 5).

Discussion

Our study is the first to assess HCT readiness in girls and young women with Turner syndrome applying the TRAQ-GV-15. We collected data from 27 girls and young women with TS at three Austrian pediatric clinics and compared the results to age-matched AYA with type 1 diabetes or a childhood onset rheumatic disease.
As hypothesized, we found statistically significant lower overall TRAQ scores for AYA with TS. Primarily, lower scores in subscale 1 “Autonomy” were responsible for lower overall scores presented by AYA with TS. Here, again, Item 05 (“Do you call the doctor’s office to make an appointment?”) and item 07 (“Do you arrange for your ride to medical appointments?”), both belonging to subscale 1. This goes in line with reports from the literature on TS (15,22), emphasizing less autonomy and higher dependence on parents, especially on mothers, in girls with TS compared to normally developed girls. Item 09 (“Do you apply for health insurance if you lose your current coverage?”) is accountable for another significant difference between TS patients and controls. This item belongs to subscale 2 “Health Literacy”. In this context it must be stated, that the TRAQ-GV-15 pilot study showed that item 09 achieved the second lowest mean score (msc 2,32) with the largest SD (±2.09) of all 15 TRAQ-GV items (culen TRAQ 2018). Nevertheless, scoring was significantly lower for the TS group. These results are in line with another recent study where patients with more health restraints and neurocognitive impairments showed lower scores (23). Higher indicators for transition readiness were found in patient groups diagnosed with a chronic physical condition without neurocognitive distinctiveness whereas lower indicators for HCT readiness were found in cohorts with cognitive and behavioral shortcomings, coming along with the primary chronic condition (23).

Differences in handling the TRAQ-GV-15 were apparent regarding to *fill-in time*. AYA of the TS group needed significantly more time to fill out the questionnaire. Reduced working speed could eventually result from less efficient information processing and impaired working memory, frequently observed in the TS literature (24). AYA with TS used significantly more *consultation time* than controls. This fact, on the one hand,

could indicate the need of intensified personal attentiveness. On the other hand, this result may be a bias due to the research focus on TS in this work. Regarding *help needed*, participants of the TS group did not need help significantly more often than controls. Nevertheless, TS patients consumed more time for discussion and for personal support pre- and post-filling-out the questionnaire.

Autonomy and reports of more discussion proved to be important for the development of transition readiness (25). Still, recent research revealed that less than 50% of AYA report discussions on the topic of HCT with their providers (12). In this study, we did not systematically collect data if conversation on transition had taken place before. However, we hypothesized that eventually the higher number of hospital visits in our control group might have entailed more personal talks with providers. This might have enhanced disease management competencies and therefore attribute to higher transition readiness scores. At any rate, surveyed YSHCN enjoyed extra talks an HCT. Therefore, clinical value might derive from the held conversations in the course of this study.

Concerning language skills, we are confident to claim language skills did not account for differences in TRAQ scores. Sufficient German language skills not only were an inclusion criteria but also more than 80% of the TS patients stated German as their first language.

Though literature documents learning difficulties in TS girls (26), reported school types did not show obvious differences between the study groups. This is in line with earlier reports (27). However, data on patient characteristic are not fully reported, thus, our results in this regard could not be tested for significance.

As all the participants in the TS study group were adequately substituted with oestrogens and the pubertal stage was comparable to the female individuals in the control group, we could conclude that the discrepancies in TRAQ scoring in the TS group cannot be explained by differences in pubertal development. Other factors, such as decreased self-organization or a lower degree of autonomy in this particular group (16), might explain these results

Notwithstanding, differences in TRAQ scores might also emerge due to differences in standard of care by different provider teams. Future research should systematically differentiate between the influence of structural, individual and conditional differences.

Strengths: This is the first study to assess transition readiness in German speaking girls and young women with TS and to compare them with an age-matched control group with chronic conditions other than TS.

Limitations: We have to state several limitations. First, this study was cross-sectional and the study population was a convenience sample. Results cannot be generalized and should be interpreted with caution. Second, sociodemographic data was sparsely documented. Therefore, the only variable accounting for differences in transition readiness in this study was the primary diagnosis. This might be a bias. Further, no supplementary questionnaire was applied to validate TRAQ-GV-15 results. Last, self-assessment always holds the risk of a bias. However, caregivers and health-care providers were not included in the study. Their influence and support for AYA is stated to be of major impact on transition outcome (28)). Therefore, these players should be included in future studies.

Conclusion:

Patients with TS seem to be less ready for transition than age-matched patients with less impairing chronic conditions. Assessing transition readiness may help in identifying underdeveloped skills or gaps in knowledge concerning disease management. The use of transition readiness assessment questionnaires in routine standard care of YSHCN could be useful for process-based preparation for transfer, for long-term follow-ups or for transition outcome research. Clinical value might emerge from detecting knowledge gaps and recognizing the lack of competencies. Additional benefit might arise from standardized discussions on the topic of transition between patient, caregivers and HCP, including the valuable opportunity of addressing uncertainties and outstanding issues. We are convinced that this approach will raise the chance of better HTC preparation for young patients with TS.

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Less ready for adulthood? – Turner syndrome has an impact on transition readiness

Figure 1: recruitment of participants for study group

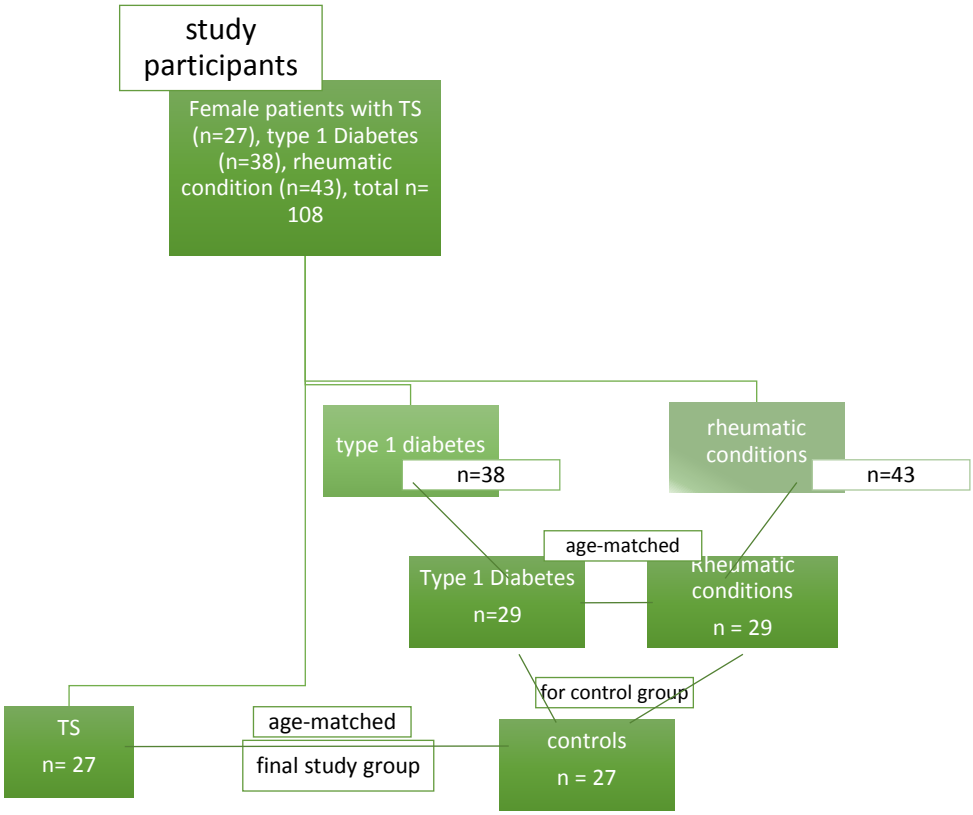

Table 1: Mean age and standard deviation from mean age in girls and young women with Turner syndrome (TS) and their age-matched controls.

	Mean ±*SD*	Min	Max
TS	17.43 ±2.28	14.49	22.56
Controls	17.60 ±2.60	14.75	23.39

Table 2: TRAQ-GV-15 Overall Mean Scores and Subscale Mean Scores for TS group and controls

	TRAQ-GV-15 total score * *p* 0.02	Domain 1 *Autonomy* ** *p* 0.009	Domain 2 *Health Literacy* *p* 0.109	Domain 3 *Adherence* *p* 0.679
Item level	Mean ±*SD* (Md)	Mean ±*SD* (Md)	Mean ±*SD* (Md)	Mean ±*SD* (Md)
TS	3.65 ± 0.64 (3.53)	3.81 ± 0.88 (3.86)	2.47 ±1.01 (2.25)	4.52 ± 0.59 (4.75)
Controls	3.95 ± 0.68 (3.73)	4.23 ± 0.77 (4.57)	2.91 ±1.28 (2.75)	4.53 ± 0.64 (4.75)
Total score level				
TS	54.70 ± 9.57 (53.0)	26.70 ± 6.19 (27.0)	9.86 ± 4.03 (9.0)	18.07 ± 2.35 (19.0)
Controls	59.23 ± 10.27 (56.0)	29.58 ± 5.38 (32.0)	11.63 ± 5.13 (11.0)	18.11 ± 2.55 (19.0)

TRAQ = Transition Readiness Assessment Questionnaire, GV= German version, SD = standard deviation, Md=median

* *p*-value of < 0.05

***p*-value of < 0.01

Table 3: TRAQ-GV-15 Single Item Mean Score for TS group and Controls

	controls			TS			
Single items TRAQ	n	Mean	*SD*	n	Mean	*SD*	*p*
item 01 (dom. 1)	27	3.56	1.63	27	3.41	1.34	0.791
item 02 (dom. 2)	27	3.78	1.48	26	3.31	1.59	0.282
item 03 (dom. 3)	26	4.77	0.71	27	4.81	0.62	0.832
item 04 (dom. 1)	26	3.85	1.46	27	3.52	1.58	0.463
item 05** (dom. 1)	27	3.96	1.32	27	3.04	1.48	0.003
item 06 (dom. 3)	27	4.41	0.97	27	4.30	1.07	0.769
item 07** (dom. 1)	27	4.15	1.32	27	3.19	1.55	0.004
item 08 (dom. 3)	27	4.26	1.16	27	4.41	1.05	0.771
item 09* (dom. 2)	27	2.67	1.64	27	1.85	1.20	0.035
item 10 (dom. 2)	27	2.56	1.67	27	2.19	1.21	0.306
item 11 (dom. 2)	27	2.63	1.55	27	2.56	1.42	0.678
item 12 (dom. 1)	27	4.70	0.82	27	4.56	0.89	0.323
item 13 (dom. 1)	27	4.48	0.89	27	4.19	1.11	0.307
item 14 (dom. 3)	27	4.67	0.56	27	4.56	0.97	0.742
item 15 (dom. 1)	27	4.85	0.36	27	4.81	0.40	0.705

TRAQ = Transition Readiness Assessment Questionnaire, SD = standard deviation, dom. = domain; Domain 1 = Autonomy, Domain 2 = Health Literacy, Domain 3 = Adherence

* *p*-value of < 0.05

***p*-value of < 0.01

Table 4: Handling of the Administration of the TRAQ-GV-15

	* Fill-in Time *p* 0.01			** Consultation Time *p* <0.001			Help Needed p 0.393		
	within 5 min	6-10 min	11-20 min	≤ 15 min	16-30 min	31-60 min	0-1 times	2-3 times	≥ 4 times
TS	54.2%	29.2%	16.7%	8.0%%	64.0%	28.0%	76.0%	16.0%	8.0%
Controls	85.2%	14.8%	0%	85.2%	14.8%	0%	85.2%	11.1%	3.7%

TRAQ = Transition Readiness Assessment Questionnaire, GV= German version
* *p*-value of < 0.05
**p*-value of < 0.01

Table 5: Data of the Study Sample on number of hospital visits, first language of the participants and educational level

		hospital visits		first language			school type			
		1-2 /y	≥ 3/y		German	others		High school	Apprentice-ship	Acadamic training
	n			n			n			
TS	22	72.7%	27.3%	27	83.3%	16.7%	24	58.3%	16.7%	25%
controls	19	10.5%	89.5%	27	66.7%	33.3%	19	84.2%	15.8%	0%

y = years
n = total number of participants who provided requested data

CHAPTER THREE: DISCUSSION AND CONCLUSION

Given the broad range of the field of transition, this work aims at providing an overview on the current status of the research with a special focus on transition readiness and the assessment of transition readiness. This research pursuit led to the development of a pilot-tested assessment instrument in German language, the TRAQ-GV-15 (Culen et al., 2019).

HCT holds extra hurdles for endocrine conditions (Bachelot et al., 2017; Paepegaey et al., 2018) and transition outcomes in patients with TS proved to be poor (Devernay et al., 2009; Ertl et al., 2018). Therefore, my work included the investigation of psychosocial care for girls with TS (Culen, Ertl, Schubert, Bartha-Doering, & Haeusler, 2017) and the comparison of transition readiness scores between patients with TS and patients with type 1 diabetes or a rheumatic condition, assessed with the TRAQ-GV-15 (Culen et al., n.d.).

The TRAQ-GV-15 is the first generic transition readiness assessment tool in German language, as far as we know. Psychometric properties of the German version are reported and reference scores regarding age and sex are available (Culen et al., 2019). The questionnaire has become an independent tool, deviating from the TRAQ 5.0 in several ways, including the reduction from 20 items to 15 items and from five subscales to three subscales. The changes in the factor structure were anticipated by the authors of the original tool (Sawicki et al., 2011). A reduction of items was suggested by the authors of the TRAQ 5.0 (Wood et al., 2014).

Cross-cultural adaptation to other languages and cultural environments showed that the instrument always underwent specific adjustments. None of the adapted tools reached the overall Cronbach´s alpha of 0.93 in the first version of the TRAQ (Sawicki et al., 2011) as well as the Cronbach´s alpha 0.94 of the TRAQ 5.0 (Wood et al., 2014). Adaptations into Brazilian Portuguese (overall Cronbach´s alpha 0.78) (Anelli et al., 2018), into Argentinian Spanish (overall Cronbach´s alpha 0.81) (Gonzalez et al., 2017) and into Turkish (overall Cronbach´s alpha 0.88) (Kızıler et al., 2018a) reported values comparable to the overall Cronbach´s alpha 0.82 of the German version (Culen et al., 2019).

Additionally, the structure of the different versions of the TRAQ differed regarding to the number of subscales and items. The first version comprised 29 items (Sawicki et al., 2011), whereas the TRAQ 5.0 (Wood et al., 2014) was reduced to 20 items. The Brazilian version as well as the Turkish version held on to the 20 items of the TRAQ. In contrast, the Argentinian version dropped the subscale “Talking to Providers, comprising two items, and resulted in an 18 items questionnaire. Following the suggestion of the authors of the TRAQ 5.0., the German version omitted five items, resulting in a 15 items instrument. Accordingly, the factor structure of the different TRAQ versions varies. Subscales emerge after psychometrical testing and depend among other parameters on the number of items and the composition of patient sample. Therefore, the suggested three-factor-structure of the German TRAQ is not found in the TRAQ 5.0 nor in the other adaptations which reported four subscales in the Argentinian version and five subscales in the Brazilian and Turkish version. Against that background, the question arises to what extent TRAQ scores can be compared internationally, derived from the different TRAQ versions. However, health care systems and medical services differ between countries and continents. Therefore, the need of a cross-cultural adapted instrument was given and will facilitate research in the field of transition in the German speaking countries.

In contrast to the different psychometric structures of the various TRAQ questionnaires, the response set derived from a transtheoretical model of change was retained by all reported adaptations (DeCunto et al., 2017; Gonzalez et al., 2017; Anelli et al., 2018; Kızıler, Yıldız, & Eren Fidancı, 2018b; Culen et al., 2019). Results were not interpreted with regard to the theoretical background of the answer options but interpreted solely regarding the Likert-scale numbers one to five. In our study, equally, TRAQ scores were taken for quantitative results. This might include loss of information as the response options comprise motivational aspects in the sense of “wanting to learn”. However, this concept may be too ambitious and too complex to be considered for interpretation of the TRAQ scores.

All cross-culturally adapted TRAQ questionnaires share the features of easy administration, good comprehensiveness and applicability for all chronic conditions (Anelli et al., 2018; Gonzalez et al., 2017; Kızıler, Yıldız, & Eren Fidancı, 2019; Culen et al., 2019).

Transition readiness scores and sex

Findings regarding TRAQ scores are inconclusive when it comes to sex and transition readiness. Some studies indicate differences between girls and boys when it comes to transition readiness, stating lower scores for boys or higher scores for girls (Sawicki et al., 2011; Wood et al., 2014; Stewart et al., 2017; Gonzalez et al., 2017). Girls scored higher in the self-advocacy scale (Sawicki et al., 2011), the medication management and the daily-activities-management scale (Wood et al., 2014). Other studies reported no differences in TRAQ scores between sexes (Anelli et al., 2018). TRAQ-GV-15 scores in our cohort showed no differences between male and female participants (Culen et al., 2019).

Transition readiness scores and age

The results of international studies using a TRAQ version are clear regarding age in that way that older age correlated positively with higher scores (Sawicki et al., 2011; Wood et al., 2014; Gonzalez et al., 2017; Anelli et al., 2018; Kızıler et al., 2018b). In our cohort, significant differences in the TRAQ-GV-15 scores were found between the younger participants aged fourteen and fifteen and the participants older than sixteen. However, as transition readiness scores showed no difference between the age of sixteen or seventeen and older than eighteen, solely chronical age will not be sufficient as a discriminant variable for transition readiness in patients due to transfer. Assessment remains reasonable and helpful.

Transition readiness scores and potential predictors

Ecological factors such as geographic area, median income, language and sex composition predicted transition readiness scores. In a recent study the UNC TRxANSITION Scale for assessing transition readiness and disease self-management was used (Javalkar et al., 2016). Again, age and female sex predicted higher scores whereas race had no influence after adjusting for ecological factors like the ZIP code. In my work, the focus was primarily on the psychometric qualities of the assessment questionnaire. Therefore, ecological factors were not taken into account except for language and educational level which showed no significant correlation with TRAQ-GV-15 scores. However, a different research question might also reveal ecological

parameters influencing transition readiness of YSHCN in the German speaking countries.

Plans for transition

A low level on transition planning and thinking about transitioning by YSHCN and their parents was reported earlier (Sawicki et al., 2014). This was confirmed during the interviews with our patient cohort. From experience with the participants in our study we realized that the majority was not aware of transitional demands. Some of the AYA were not even aware of the need to transfer. This goes in line with the literature which pinpoints that YSHCN are not adequately prepared for transfer (Jensen et al., 2017). A limitation of our study is the lack of documentation on earlier discussions about transition with health care providers or parents and on having plans for transition. However, the consciousness for the importance of transition issues is certainly rising. Parents and caregivers articulated the need for HCT services for patients with neurocognitive conditions, such as ASD (Kuhlthau, Delahaye, Erickson-Warfield, Shui, & Crossman, 2016) Transition in the mental health field proves to be equally challenging (the MILESTONE Consortium et al., 2018).

In my experience AYA and parents wish discussions and individualized information on transitional issues, preferably with people they have a relationship with (Freeman, Stewart, Cunningham, & Gorter, 2018). AYA showed a high degree of acceptance when difficult and intimate issues relevant for transition were addressed. I am convinced that the process of answering the items of the TRAQ-GV-15 itself triggered awareness for transition issues such as autonomous disease management and raised questions regarding future health care and areas in need of development. Furthermore, conversations about transition between AYA and parents were initiated. Earlier findings undermine these observations (Williams et al., 2010).

Transition readiness scores and TS

In the course of the international cross-cultural adaptations of the various TRAQ versions, reports on condition related TRAQ scores were not included in the research scope. The aim at designing a generic tool, applicable to all kind of patient cohorts, retreated this question in the background (Anelli et al., 2018; Wood et al., 2014; Kızıler et al., 2018b; Culen et al., 2019). However, subsequent studies using the TRAQ

indicated that a specific chronic diagnosis refers as a reliable variable to significantly different TRAQ scores (Beal et al., 2016).

After pilot-testing the TRAQ-GV-15, I was interested in potential differences in TRAQ scoring between our TS patient cohort and other chronic conditions not known for a comparable complex clinical picture comprising psychosocial and cognitive impairments. Findings indicate lower scores for conditions associated with cognitive impairments (Sawicki et al., 2011; Beal et al., 2016), which might demand special attention to chronic conditions such as TS, associated with the risk for neurocognitive impediments.

No significant differences in the TRAQ-GV-15 scores were observed between girls and young women with diabetes or a rheumatic condition. In contrast, TRAQ-GV-15 scores of girls and young women with TS differed significantly in several aspects. First, TS patients scored significantly lower in subscale 1, which is subtitled with the term “autonomy”. This reflects descriptions of girls with TS as being less autonomous than age-matched peers (J. L. Ross et al., 2002; David S. Hong et al., 2011). Even more detailed information is available when looking on the item-level differences. Two items that ask for independent behavior such as arranging a medical appointment and organizing the ride to the doctoral visits showed significant lower scores. Reports from the literature support explanations such as TS patients being less autonomous or their parents being more deeply involved into the lives of their daughters (Gawlik et al., 2012; Downing et al., 2013). Promoting autonomy in patients is regarded as an ethical issue and has been related to increased wellbeing (the MILESTONE Consortium et al., 2018). These findings should be encouraging to support autonomous conduct in AYA with TS and to gradually decrease paternalism in order to enhance transition readiness skills in girls with TS.

Decreased working speed and limited working memory have been recognized for TS (M. M. M. Mazzocco, 2006; D. Hong et al., 2009). In our findings, participants with TS needed significantly more time to fill out the TRAQ-GV-15. This might indicate that AYA with TS need more time to grasp relevant contents or to make decisions. However, since patients with TS did not need more help in filling out the questionnaire, it might be sufficient to provide more time than usually scheduled in time-restricted doctoral visits.

Since HCT coincidences with an important developmental phase of AYA including pubertal development, hormonal aberrations, fertility, first experiences with love life and growing interest in sexual issues, I saw the need to meet AYA without their medical doctors, parent or otherwise caregivers such as grandparents or older siblings. are more relevant. Therefore, information on parental involvement and the caregiver's perspective is absent. No triangulation of my findings with patients, parents and health experts was possible. A different study design might overcome these methodological shortcomings.

Transition readiness assessment implementation

The area "transition readiness" improved the most within an one-year observation period after implementation according to an evaluation of transition services (McManus, Ilango, Beck, & White, 2017; Ilango, Beck, McManus, & White, 2018) within the six core elements concept ("GotTransition.org," 2016).

This might be due to the fact that policy implementation on a systemic level and tracking transition completion with long-term follow-ups are more complex tasks than assessing transition readiness in individuals. Therefore, we are encouraged to follow the strategy of promoting and facilitating the assessment of transition readiness in our clinic. The clinical use of the TRAQ questionnaire is well underway internationally, e.g. as baseline assessment for comprehensive research (Beal et al., 2016) or disease specific transition programs (Mackie et al., 2014; Little, Odiaga, & Minutti, 2017; Carlsen et al., 2017). Cross-cultural adaptations (Kızıler et al., 2019) demonstrate the great interest in validated assessment tools. Revisions from earlier developed instruments (Moynihan, Saewyc, Whitehouse, Paone, & McPherson, 2015) manifest the ongoing efforts in improving questionnaires used in the transition process.

Transition services and next steps

Recommendations for the clinical work include the implementation of transition case managers or personnel in charge of transitional issues, transition consultation hours or joint office hours. Inadequate transition preparation in YSHCN emphasizes the need also for HCP to better prepare YSHCN for transition (Lebrun-Harris et al., 2018). In preparation of transition services, infrastructure to facilitate collaboration and an online data repository are needed (Gray, 2018). The evaluation of the Ready Steady

Go-program (Nagra et al., 2015) encourages to follow the path of improving transition services.

Websites of the pediatric hospitals are increasingly providing comprehensive information regarding transition including download materials and helpful links, designed to support patients, caregivers and HCP. This would also be preferable for Austrian pediatric clinics.

The Society for Transition Medicine ("Deutsche Gesellschaft für Transitionsmedizin," 2018) plans to provide overarching information, tools, helpful links, videos featuring testimonials and the dissemination of new collaborative efforts such as assisted housing or holiday camps in cooperation with sponsors, politics and industry.

Future research

Future research regarding the TRAQ-GV-15 should include an evaluation of the subscale structure, conducting Confirmatory Factor Analysis on a larger convenience sample.

It would be of great interest, whether TRAQ scores are influenced by the current response options. No study investigated this question as far as we know. A randomized controlled study with two different response sets could clarify this question.

Future deployment of the TRAQ-GV-15 should involve caregivers for several reasons. Parental involvement increases transition readiness in YSHCN. On the other hand, enhanced autonomy for AYA means detachment from the parents. This process will develop more smoothly when caregivers are prepared and engaged. In addition, the association of TRAQ-GV-15 scores according to experts' and caregivers' assessment is needed to investigate face validity. Triangulation of findings will identify areas of consistent estimates and different understandings.

Longitudinal studies will test the predictive validity of the TRAQ-GV-15. Ideally, the administration of the TRAQ-GV-15 will be embedded in a structured transition program. In that case, evaluation of the instrument will have to be judged in that context. As younger patients are more likely to complete follow-up assessment questionnaires (Calhoun et al., 2019), this information emphasizes the administration of the TRAQ in early adolescence and indicates a better chance for prospective data.

Linking TRAQ-GV-15 scores to medical data might also be helpful in interpreting the predictive validity of the scores.

Therefore, a follow-up of the participants of our reported studies would be desirable. Assessing differences in the TRAQ-GV-15 scores two or three years after baseline assessment would be of great value. Additionally, investigation on whether discussions on transitional issues have been addressed either from YSHCN, from parents or health experts subsequently to the study or not would be of great interest.

In the hope of finding parameters that are modifiable it is of great importance to investigate ecological predictors of transition readiness in German speaking countries. On a system level, risks for failed transition could be detected and prevention strategies or at least awareness on a structural level could be developed. Socioeconomic status assessed by occupational life of the parent or ZIP code, success in school assessed by school grades or sociodemographic data including family composition or living situation could reveal correlations with transition readiness.

Other predictors for transition readiness such as self-efficacy, resilience or autonomy or plans-for-the-future have been investigated in earlier studies. However, research on these questions is lacking for German speaking YSHCN. Again, finding predictors that can be enhanced by either education or training is of major interest.

Collaboration with working groups nationwide, or in the German speaking countries respectively, would provide larger patient cohorts and would facilitate research on a systemic level comprising comparison of health care transition services and transition programs internationally. Surveying satisfaction of patients and caregivers with transitional services might be facilitated by the disease neutral TRAQ-GV-15.

Next developmental steps may lead to an online version of the TRAQ-GV-15, making it easier for YSHCN to fill out the questionnaire remotely after having received a link on their mobile device. Equally for HCP, digital application and online documentation of the TRAQ-GV-15 might reduce workload and simplify administration processes.

Conclusion

With this research, I wanted to facilitate the incorporation of an applicable and time-efficient transition readiness assessment tool in clinical routine care. The TRAQ-GV-15 is easy to administer, feasible and economic. Since the need for improved transition processes and for transition readiness assessment is ubiquitous and validated tools for assessment were lacking in the German language, I am confident that the TRAQ-GV-15 will find acceptance in the German speaking world of health care services. With this work, I hope I can substantially contribute to better preparing German speaking AYA with chronic conditions for transition.

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APPENDIX

Curriculum Vitae

Mag.[a] Caroline Culen
Geboren am 1. April 1971 in Wien
Verheiratet, vier Kinder (1994; 1997; 2005 und 2007)

Ausbildung

1974-1975	französischer Kindergarten, Fontainebleau, Frankreich
1977-1981	Volksschule Maria Regina in Wien
1989	Matura (neusprachlich) am Gymnasium Maria Regina in Wien
9/1989-2/1990	Französischsprachkurs in Tours, Frankreich; Arbeitspraktikum in Paris
1990-1997	Studium der Psychologie an der Universität Wien
1993	6-Wochenpraktikum an der Klinik Menterschwaige, Fachklinik für Psychiatrie Psychotherapie Pychoanalyse, München
1999	Abschluss des Universitätslehrgangs „Psychotherapeutisches Propädeutikum“
1999-2002	Ausbildung „Teamsupervision und Coaching“, Berufsverband österreichischer PsychologInnen (BÖP)
1999-2004	Assistenz in der Erwachsenenbildung (Kurt Fleischner)
1999-2006	Teilnahme an Kongressen, Tagungen und Weiterbildungsseminaren in

	systemischer Beratung und Aufstellungsarbeit (u.a. Steve de Shazer, Matthias Varga von Kibéd, Bert Hellinger, Österreichisches Forum für Systemaufstellungen, ÖfS)
2006-2007	Weiterbildung „Dialogisch–systemische Aufstellungs– und Rekonstruktionsarbeit“ Apsys Graz (Ch. Essen und G. Baxa)
2008-2010	Lehrgang „Kinderschutzarbeit“, NÖ Landesakademie—NÖ Heilpädagogisches Zentrum Hinterbrühl
2010-2011	Curriculum „Klinische Psychologie und Gesundheitspsychologie“, Österreichische Akademie für Psychologie (ÖAP)
2012	Eintragung in die Liste des Bundesministeriums als Klinische (Eintragungnr: 8877) und Gesundheitspsychologin (Eintragungn: 8850)
seit 2012	laufende Fortbildungen, Seminare und Kongresse im Bereich klinische Psychologie, Gesundheitspsychologie und Psychologie des Kindes- und Jugendalters
10/ 2015	Beginn des Doctoral Programme of Applied Medical Science N790 Studiums an der Medizinischen Universität Wien, thematisches Programm „Public Health“ – Forschungsschwerpunkt TRANSITION chronisch kranker Kindern und Jugendlichen von pädiatrischer Versorgung in die Erwachsenenmedizin.

Fremdsprachen:

Englisch und Französisch in Wort und Schrift

Spanisch Grundkenntnisse

Latein Schulkenntnisse

Einschlägige berufliche Tätigkeiten

1998-1999	Psychosoziales Praktikum im Sanatorium Maimonides–Zentrum
2000	Kick Off Management Consulting GmbH
2001 – 2005	freiberuflich als Supervisorin und Coach in freier Praxis/Gemeinschaftspraxis
2005 – 2007	Karenz und Pflegekarenz aufgrund einer onkologischen Erkrankung der Tochter
2007 – 2008	Karenz 4. Kind
2008 – 2013	„die Möwe“—Kinderschutzzentren, Bereich Prävention: Entwicklung von Präventionsprojekten Vortragstätigkeit, Informations– und Fortbildungsveranstaltungen für PädagogInnen, Elementar- und HortpädagogInnen; Elternberatung und –schulung, Workshops mit Kindern und Jugendlichen
2011 – 2016	Vortragstätigkeit im Lehrgang „Kinderschutzarbeit“, NÖ Landesakademie—Heilpädagogisches Zentrum Hinterbrühl
2011 – 2012	praktische Fachausbildung zur Klinischen Psychologin an der Universitätsklinik für Kinder und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pulmologie, Allergologie und Endokrinologie, Schwerpunkt Diabetes und Cystische Fibrose
2013 – 2014	Projektleitung an der Universitätsklinik für Kinder- und Jugendheilkunde AKH Wien: Konzeption, Organisation und Durchführung von Coachingruppen für Eltern von Kindern mit T1DM), Drittmittelfinanzierung
2014	Gründung des Vereins www.cuko.care gemeinsam mit Dr.in Marianne König

2014 – 2015	Karenzstelle KAV, Abteilung für Pulmologie, Allergologie und Endokrinologie, Universitätsklinik für Kinder- und Jugendheilkunde AKH Wien
ab 2015	wissenschaftliche Mitarbeiterin im Bereich Endokrinologie an der Universitätsklinik für Kinder- und Jugendheilkunde des AKH Wien, Drittmittel, regelmäßige Vortragstätigkeit zu Themen der Kinder- & Jugendgesundheit
ab 04/2017	Leitung fachliche Stabstelle in der Österreichischen Liga für Kinder- und Jugendgesundheit, www.kinderjugendgesundheit.at
ab 11/2017	Fachliche Geschäftsführung Österreichische Liga für Kinder- und Jugendgesundheit
ab 2019	Geschäftsführung Österreichische Liga für Kinder- und Jugendgesundheit

Comparison TRAQ 5.0 and TRA-GV-15 Questionnaires

TRAQ – Transition Readiness Assessment Questionnaire

TRAQ 5.0 (Wood 2014)	TRAQ-GV-15	New scales, TRAQ-GV-15
Managing Medications	**Medikamente**	**Selbständigkeit/Autonomy**
1. Do you fill a prescription if you need to?	1. Kannst Du selbst Rezepte einlösen?	5. Vereinbarst du Arzttermine selbst?
2. Do you know what to do if you are having a bad reaction to your medications?	2. Weißt Du, was im Fall von Nebenwirkungen Deiner Medikamente zu tun ist?	1. Löst Du selbst Rezepte ein?
3. Do you take medications correctly and on your own?	3. Nimmst Du selbständig Deine Medikamente regelmäßig und nach Vorschrift?	4. Kümmerst Du Dich um die rechtzeitige Nachbestellung Deiner Medikamente?
4. Do you reorder medications before they run out?	4. Kümmerst Du Dich um die rechtzeitige Nachbestellung Deiner Medikamente?	7. Organisierst Du Deine Fahrt zu Kontrollterminen selbst?
Appointment Keeping	**Arzttermine**	12. Füllst Du medizinische Fragebögen selbst aus?
5. Do you call the doctor's office to make an appointment?	5. Vereinbarst du Arzttermine selbst?	13. Führst Du händisch/elektronisch eine Liste mit Deinen Terminen?
6. Do you follow-up on any referral for tests, check-ups or labs?	6. Folgst Du Empfehlungen zu weiteren speziellen Untersuchungen oder Labortests?	**Gesundheitskompetenz/Health Literacy**
7. Do you arrange for your ride to medical appointments?	7. Organisierst Du Deine Fahrt zu Kontrollterminen?	10. Weißt Du, welche Leistungen Deine Krankenversicherung bezahlt?
8. Do you call the doctor about unusual changes in your health (For example: Allergic reactions)?	8. Meldest Du Dich bei gesundheitlichen Problemen (z.B. allergischen Reaktionen, etc..) bei einer Ärztin/einem Arzt?	11. Übernimmst Du selbst Geldangelegenheiten und Lebenshaltungskosten?
9. Do you apply for health insurance if you lose your current coverage?	9. Kümmerst Du Dich um Deine Kranken-versicherung, falls Deine Mitversicherung endet?	9. Kümmerst Du Dich um Deine Krankenversicherung, falls z.B. die Mitversicherung bei Deinen Eltern endet?
10. Do you know what your health insurance covers?	10. Weiß Du, was Deine Krankenversicherung alles bezahlt?	2. Weißt Du, was im Fall von Nebenwirkungen Deiner Medikamente zu tun ist?
11. Do you manage your money & budget household expenses (For example: use checking/debit card)?	11. Verfügst Du über eigenes Geld und übernimmst Du auch Lebenshaltungskosten?	**Adhärenz/Adherence**

TRAQ – Transition Readiness Assessment Questionnaire

Tracking Health Issues	**Gesundheitsbezogene Fragen**	14. Teilst Du Ärztinnen/Ärzten oder Pflegepersonal mit, wie Du Dich fühlst?
12. Do you fill out the medical history form, including a list of your allergies?	12. Füllst Du medizinische Fragebögen selbst aus?	6. Folgst Du Empfehlungen zu weiteren speziellen Untersuchungen oder Labortests?
13. Do you keep a calendar or list of medical and other appointments?	13. Führst Du händisch oder elektronisch eine Liste mit Deinen Terminen?	3. Nimmst Du Deine Medikamente nach Vorschrift und selbständig?
14. Do you make a list of questions before the doctor's visit?	....	8. Meldest Du Dich bei gesundheitlichen Problemen (z.B. allergischen Reaktionen, etc..) bei einer Ärztin/einem Arzt?
15. Do you get financial help with school or work?	....	15. Beantwortest Du Fragen, die Dir das medizinische Personal stellt selbst?
Talking with Providers	**Gespräche mit medizinischem Personal**	
16. Do you tell the doctor or nurse what you are feeling?	14. Teilst Du Ärztinnen/Ärzten oder Pflegepersonal mit, wie Du Dich fühlst?	
17. Do you answer questions that are asked by the doctor, nurse, or clinic staff?	15. Beantwortest Du die Fragen, die Dir das medizinische Personal stellt.	
Managing Daily Activities	....	
18. Do you help plan or prepare meals/food?	....	
19. Do you keep home/room clean or clean-up after meals?	....	
20. Do you use neighborhood stores and services (For example: Grocery stores and pharmacy stores)?	....	

TRAQ_DV_cc_Version 2.1 April 2017

Fragebogen zur Transitionsbereitschaft (TRAQ –Transition Readiness Assessment Questionnaire)

Anweisungen für Jugendliche und junge Erwachsene: Bitte kreuze die Kästchen an, die Deine Fähigkeiten am besten beschreiben. Es gibt keine richtigen oder falschen Antworten. Deine Antworten werden vertraulich behandelt. Bitte hier ankreuzen, falls Du den Fragebogen selbst ausgefüllt hast: ☐

Anweisungen für Eltern/Erziehungsberechtigte: Falls Ihre Tochter/Ihr Sohn die untenstehenden Fragen nicht alleine beantworten kann, bitten wir Sie stattdessen jene Kästchen anzukreuzen, die Ihre eigenen Fähigkeiten in diesen Bereichen am besten beschreiben. Bitte hier ankreuzen, falls Sie den Fragebogen ausgefüllt haben: ☐

Studiencode: Heutiges Datum: _____.____.____	Nein, da kenne ich mich nicht aus	Nein, ich möchte aber gerne dazulernen	Nein, aber ich bin gerade dabei es zu lernen	Ja, manchmal	Ja, immer wenn nötig	1-5
Medikamente						
1. Löst Du selbst Rezepte ein?						
2. Weißt Du, was im Fall von Nebenwirkungen Deiner Medikamente zu tun ist?						
3. Nimmst Du Deine Medikamente nach Vorschrift und selbständig?						
4. Kümmerst Du Dich um die rechtzeitige Nachbestellung Deiner Medikamente?						
Arzttermine						
5. Vereinbarst du Arzttermine selbst?						
6. Folgst Du Empfehlungen zu weiteren speziellen Untersuchungen oder Labortests?						
7. Organisierst Du Deine Fahrt zu Kontrollterminen selbst?						
8. Meldest Du Dich bei gesundheitlichen Problemen (z.B. allergischen Reaktionen, etc..) bei einer Ärztin/einem Arzt?						
9. Kümmerst Du Dich um Deine Krankenversicherung, falls z.B. die Mitversicherung bei Deinen Eltern endet?						
10. Weißt Du, welche Leistungen Deine Krankenversicherung bezahlt?						
11. Übernimmst Du selbst Geldangelegenheiten und Lebenshaltungskosten?						
Gesundheitsbezogene Fragen						
12. Füllst Du medizinische Fragebögen selbst aus?						
13. Führst Du händisch/elektronisch eine Liste mit Deinen Terminen?						
Gespräche mit medizinischem Personal						
14. Teilst Du Ärztinnen/Ärzten oder Pflegepersonal mit, wie Du Dich fühlst?						
15. Beantwortest Du Fragen, die Dir das medizinische Personal stellt, selbst?						
be on TRAQ DV_2.1 c. culen, März 2017 Korrespondenz: caroline.culen@meduniwien.ac.at						

TRAQ-GV-15

TRAQ GV-15, Transitionsbereitschaftsfragebogen

Alle unten angeführten Bereiche sind für den Übergang, die Transition, in die Erwachsenenbetreuung wichtig. Es gibt keine richtigen oder falschen Antworten. Die Antworten werden vertraulich behandelt.

Anweisungen für Jugendliche und junge Erwachsene:
Bitte kreuze unten diejenigen Kästchen an, die Deine Fähigkeiten aus Deiner Sicht am besten beschreiben.

Heutiges Datum: _____ - _____ - _____ **Name:** **Geburtsdatum:** _____ - _____ - _____	**Nein, da kenne ich mich nicht aus**	**Nein, ich möchte aber gerne dazulernen**	**Nein, aber ich bin gerade dabei es zu lernen**	**Ja, manchmal**	**Ja, immer wenn nötig**	**Score**
1. Löst Du selbst Rezepte ein?						
2. Weißt Du, was im Fall von Nebenwirkungen Deiner Medikamente zu tun ist?						
3. Nimmst Du Deine Medikamente nach Vorschrift und selbständig?						
4. Kümmerst Du Dich um die rechtzeitige Nachbestellung Deiner Medikamente?						
5. Vereinbarst du Arzttermine selbst?						
6. Folgst Du Empfehlungen zu weiteren speziellen Untersuchungen oder Labortests?						
7. Organisierst Du Deine Fahrt zu Kontrollterminen?						
8. Meldest Du Dich bei gesundheitlichen Problemen (z.B. allergischen Reaktionen, etc..) bei einer Ärztin/einem Arzt?						
9. Kümmerst Du Dich um Deine Krankenversicherung, falls z.B. die Mitversicherung bei Deinen Eltern endet?						
10. Weiß Du, welche Leistungen Deine Krankenversicherung bezahlt?						
11. Übernimmst Du selbst Geldangelegenheiten und Lebenshaltungskosten?						
12. Füllst Du medizinische Fragebögen selbst aus?						
13. Führst Du händisch/elektronisch eine Liste mit Deinen Terminen?						
14. Teilst Du Ärztinnen/Ärzten oder Pflegepersonal mit, wie Du Dich fühlst?						
15. Beantwortest Du Fragen, die Dir das medizinische Personal stellt, selbst?						
© TRAQ 5.0 Wood, Sawicki, Reiss, Livingood &Kraemer, 2014 © TRAQ GV-15 Culen, Wood, Straub, 2019					**Total:**	

Medizinisch-Wissenschaftlicher Fonds des Bürgermeisters der Bundeshauptstadt Wien

Medizinisch-Wissenschaftlicher Fonds
des Bürgermeisters der Bundeshauptstadt Wien
Geschäftsstelle: 1030 Wien, Thomas-Klestil-Platz 8, 6. Stock, CB 16.616
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MA 40 – GMWF 506665/2016 Wien, 03. November 2016
Ihr Ansuchen auf Gewährung von Fördermitteln

Dr.in Gabriele Haeusler
c/o AKH Wien & Medizinische Universität Wien,
Universitätsklinik für Kinder und Jugendheilkunde,
Klinische Abteilung für Pulmologie, Stoffwechsel und Endokrinologie
Währinger Gürtel 18-20///
1090 Wien

Sehr geehrte Frau Dr.in Haeusler,

das Kuratorium des "Medizinisch-Wissenschaftlichen Fonds des Bürgermeisters der Bundeshauptstadt Wien" hat in seiner Sitzung am 03. November 2016 beschlossen, Ihnen für die Durchführung Ihrer wissenschaftlichen Forschungsarbeit:

Projektnummer:	15203
Projekttitel:	Be on TRAQ — Übersetzung und Anpassung des TRAQ Fragebogens zur Erhebung von Bereitschaft zur Transition, zur Anwendung bei deutschsprachigen Jugendlichen mit chronischer Erkrankung (YSHCN)

einen Betrag von € 15.000,00 aus den Mitteln des Fonds zu gewähren (die endgültige Verteilung des Förderbetrags in Personal- und/oder Sachaufwand obliegt Ihnen in Ihrer Eigenschaft als Projektleiter/in und ist im Rahmen der Vertragsunterzeichnung bekannt zu geben).

Verkehrsverbindung: Station Erdberg, erreichbar: Linien U3, Ausgang: Nottendorfer Gasse / Schnirchgasse
DVR: 0000191 -V515

Certificate Highly Read Author

Transition Award 2018

Transitionspreis 2018

17. November 2018

Beim 7. Kongresses der Gesellschaft für Transitionsmedizin e.V. wurden am am 17. November 2018 folgende Preise verliehen:

- **Caroline Culen** (Wien) für ihr Poster mit dem Thema: Be on TRAQ – Weiterentwicklung der deutschen Version TRAQ-GV-15 in Bezug auf Skalenstruktur und Reliabilität sowie Ergebnisse der ersten Pilotstudie
- **Sebastian Freilinger** (München) für sein Poster mit dem Thema: Pulmonale Hypertonie bei Erwachsenen mit angeborenen Herzfehlern (EmaH), Versorgungssituation, Wissensstand und spezifischer Beratungsbedarf
- **Andre Neutag** (Görlitz) für seinen Vortrag und Engagement im Rahmen „Junge DGM“: Selbsthilfe und Jungendarbeit für eine aktive Beteiligung junger Muskelkranker

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